

# *Pediatric Surgery*

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## **INTRODUCTION TO PEDIATRIC SURGERY**

Welcome to your Pediatric Surgery rotation. This handbook will be a guide for your time on our service. As pediatric surgery is a growing and evolving field, the pathways and treatment options described here may change over time, but the information offered will serve as a reasonable starting point. The goal of this rotation is to provide a rich and unique educational experience in pediatric surgery. For some, it may be the beginning of a career in our discipline. For those who are drawn to other areas of surgery, we hope that your time on the service will be enlightening and will help to round out your surgical educational experience while perhaps providing some unique skills and insights that may serve you in your future field.

We echo the overall principles of the Brown General Surgery Program by stating that teamwork and respect for patients, staff, and one another are fundamental for the care of children. We welcome any feedback that will help to improve our service and the educational experience of future rotating trainees.

## **ROLES, RESPONSIBILITIES, AND GOALS**

### **Fellow**

The fellow is the keystone of the service. All information should flow through the fellow, both up and down the chain of command, and the fellow is responsible for ensuring the smooth working of the service. He/she is also the face of the Pediatric Surgery Division and is responsible for interacting and communicating with staff from nursing and other services participating in the care of pediatric surgery patients.

#### Roles and responsibilities:

- To oversee all aspects of the daily functioning of the service
- To round in the NICU and PICU and to oversee the work of the PGY-1 on the floor
- To develop a daily plan for all patients seen by the service
- To oversee all consults performed by the service
- To scrub on all index cases, often with the PGY-3 as a co-scrub
- To organize the care of all patients going to the OR and to ensure their pre-operative preparation is complete (e.g. consent, booking, prep)
- To advise and educate the medical students, PGY-1, and PGY-3
- To organize weekly conference
- To present and/or supervise presentation at bi-weekly M&M
- To present at bi-weekly Pediatric Surgery conference or to arrange for a speaker for this conference
- To attend Fellow's Clinic once a week
- To organize ECMO consults, participate in cannulation, and help to direct patient care while cannulated
- To respond to all Level A and B trauma activations, and Level C activations as needed by the junior on the service
- To act as the surgical team leader in the trauma room

#### Goals:

- To have, at the completion of a two-year fellowship, the skills and knowledge needed for the independent provision of pediatric surgery care

### **PGY-3**

The duties and responsibilities of the PGY-3 overlap significantly with those of the fellow. During times when the fellow is away or on an off-service rotation (such as NICU or urology) the PGY-3 will assume the responsibilities as described above for the fellow.

#### Roles and responsibilities:

- To oversee the PGY-1s in completion of floor work and notes
- To oversee the PGY-1 when seeing floor and ED consults
- To assist and oversee the PGY-1 with bedside procedures
- To oversee and teach medical students and PGY-1s
- To help maintain the Hasbro inpatient list and the NICU list
- To round in the PICU and/or NICU, as agreed upon with the fellow
- To place all consults for PICU patients

- To perform post op checks for PICU and NICU patients
- To respond to consults placed by the PICU or NICU
- To communicate plans to the primary teams of consult patients
- To respond to all Level A and B trauma activations, and Level C activations as needed by the junior on the service
- To act as the surgical team leader in the trauma room in the absence of the fellow or attending
- To present at bi-weekly Pediatric Surgery M&M conference
- To complete at least one presentation for bi-weekly Pediatric Surgery Conference
- To coordinate communication among the members of the team (attending, fellow, APPS, and PGY-1s)
- To attend one half day of pediatric surgery clinic per week  
Normally, this clinic will be with the attending who was on-service the week prior; discuss which day is best with the attending while he/she is on service

Goals of rotation:

- To understand the physiology and care of the neonatal patient
- To understand the unique aspects of the care of critically ill pediatric general surgery and trauma patients, and how this may differ from the care of similarly ill adults
- To gain experience in the performance of pediatric cases, including pyloromyotomy and hernia repair
- To gain exposure to more complicated cases, such as esophageal atresia repair, through co-scrubbing with the fellow

## PGY-1

The PGY-1 is the hub of care for pediatric surgery patients admitted to the general floor. They are expected to manage all floor issues and to be the face of the service on the floor for both patients and other staff

Roles and responsibilities:

- To complete all daily floor-work and notes for patients on the pediatric surgery floor
- To answer all floor pages
- Ensure all tests and studies discussed in the morning are completed
- To call all consults for floor patients
- To perform post op checks for floor patients
- To complete all discharge summaries for floor patients, arrange for follow up appointments, write for home prescriptions, and arrange for home services as needed
- To round in the afternoon before sign out
- To update patient and families regarding the results of test performed and any changes in plan in a timely manner
- To maintain the floor list
- To respond to consults from the floor and ED
- To review all consults with the PGY-3/fellow
- To communicate plans to the primary teams of consult patients
- To inform the senior or fellow of any changes in patient status
- To attend and provide sign out before and after each shift
- To respond to all trauma activations
- To attend weekly Pediatric Surgery Conferences Wednesday morning

- To present once during the rotation at Pediatric Surgery conference
- To oversee and teach medical students on the rotation
- To attend Burn Clinic on Wednesday afternoons
- To attend one half day of pediatric surgery clinic per week  
Normally, this clinic will be with the attending who was on-service the week prior; discuss which day is best with the attending while he/she is on service

Goals of rotation:

- To gain a better understand of the care of the pediatric surgery patient
- To understand the physiology and care of common pediatric surgery conditions, such as pyloric stenosis and appendicitis
- To gain technical experience in the OR by scrubbing on cases such as umbilical hernia repairs and appendectomies
- To gain confidence with common procedures such as chest tube removal and abscess drainage

## PHONE NUMBERS

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### Makayla Lourenco, NP

C/P: 401-528-7905/401-221-0577

Office: 401-421-1939

Fax: 401-868-2319 or 401-444-6603

Operator 444-4000

### **Operating Rooms**

OR Desk 444-5657

Blue Room 444-6086

Preop 444-7985

PACU 444-6887

PICU 444-6021/6023

4<sup>th</sup> Floor 444-6041/6042

5<sup>th</sup> Floor 444-6051/6052

6<sup>th</sup> Floor 444-6061

### **Radiology**

Day 444-8275/Night 444-2727/4123

Ultrasound 444-0101

Body 444-4123

Sedation 444-6091

### **Emergency Department 444-4900**

Trauma Room 444-6555/6556

Express Care 444-3000

Language Line (866) 874-3972 code 229078

**NICU** 273-1122

x43300 3<sup>rd</sup> floor; x43200 2<sup>nd</sup> floor

# DAILY ROUNDS AND ROUTINE SERVICE ACTIVITIES

## I. Sign Out

- Intern AM sign out: 6am at location pre-designated by team
  - For AM sign out, the night intern will have updated, printed copies of the list ready and will review pertinent overnight events as well as new patients
  - The night intern will also obtain the I/O's and calculate the UOP (cc/kg/hr) for the day team
- Senior AM sign out: 6 am, in Fellow's office or on the go to NICU or PICU
- APP AM sign out: with seniors or at 8 AM after morning rounds
- PM: team sign out with interns and on-call senior and APPS in MOC-190 conference room

## II. NICU Rounds

- Vitals, I/O's, labs and studies should be obtained from Cerner
- Important information on AM rounds:
  - Weight and weight changes
  - Input and output (including stool), urine in cc/kg/hr and stool in cc/kg/day if ostomy present or concern for dumping
  - NGT output, including color
  - Chest tube output, presence of air leak
  - TPN: calories, access, total lipids
  - Drips
  - Amount and type of oral feeds
  - Medications
  - Results of labs and studies in last 24 hours
  - Physical exam, including appearance of bowel (gastroschisis), ostomy, wound
  - Vent settings, if applicable, or respiratory support
    - Other information can be obtained from the Results → Fellow's View tab. This includes any supplemental respiratory support, A/B/D events, what the baby is taking orally and how much and any medications the patient is on.
- When rounding in the NICU, try to speak to the overnight nurses for updates
- White coats are not to be worn in patient rooms and hands must be sanitized prior to entering each room as well as on exit
- The NICU fellow should be updated at the completion of rounds. They may be found between 6-630 in the fellow call room on the 2nd floor (code 1704#)

## III. Hasbro Rounds

### PICU

- Rounds at Hasbro typically start in the PICU with the most ill child
- Nurses should be in rounds in order to get overnight updates.
- Evaluation during rounds proceeds by systems (Neurological, respiratory, cardiovascular, GI, GU/FEN, ID, Heme, lines, etc.).

- Please note all of the issues listed above for NICU patients, as well as obtain input from the PICU service regarding their aspects of the patient's care
- Long term patients should have a discharge document started which should be updated every Friday

## Floor

- From the PICU, rounds typically start on the highest floor-highest patient
  - Exceptions are made if there are floor patients of high concern
- Effort should be made to review patients with the overnight nurses
- Please do a full exam in the morning (heart/lungs/abdomen/wound/other pertinent areas). Do not document an exam if one is not performed
- Please make note of all ins and outs, including ostomy output, NGT (including color and nature of fluid), and urine output (measures in cc/kg/hour)
  - A note about documentation: please use ONLY approved abbreviations in H&P/Consults/progress notes. A list of approved abbreviations can be found here: <http://clinical.BrownHealth.org/abbreviations>
- Please check the chart for notes from other services and have this information ready for rounds
- Please be cautious about publishing plans before discussing them with the attending or fellow. Giving other services/nursing staff only one morning plan will help to decrease confusion
- If it is anticipated that a patient will be discharged in the next 48 hours, please start a discharge summary that will be added to as needed and think proactively about medications/services/etc. that may be needed for home care
- Long term patients should have a discharge document started and the summary should be updated every Friday

## BASIC ADMISSION ORDERS

Admit order: Admit to Observation (if short stay) or admit to inpatient

IVF: D5 + LR unless otherwise specified

Diet: NPO + sips with meds unless otherwise specified; otherwise use pediatric diets

Activity: Ambulate TID unless otherwise specified

Vital signs: every 4 hours/every shift

Intake and outputs: Pediatrics/strict if specified

Children < 1 year: daily weights

Incentive spirometer: every hour

Monitoring: per attending

## LIST HANDOFF

-Brief history (does not need to include entire H&P)

-Surgery or procedure, date, attending

-**Main** To-Do items (not entire hospital course)

-Auto-populate boxes with Vitals, Diet/Drips, I&O's, POD#

-Calculations

- -UOP in cc/kg/hr
- -BM in # or stool/stoma output in cc/kg
- -Diaper mix in cc/kg
- -Emesis in # of NG output in cc/kg
- -Drain output in cc
- -Chest tube output in cc

-**Always** refresh prior to printing

## LIST MANAGEMENT

-Add consults to list as soon as you get them

-If admitted to us, should be under Pediatric Surgery

-List under admitted attending (unless someone else claims them)

-Change attending to operating surgeon once they undergo a procedure

-Update problem list and select hospital + principal problem

-Use brief, comprehensive problem names, to guide care

- Review and update Estimated Discharge Date, even if unknown
- Ensure Medication Reconciliation is complete

## **NEW CONSULTS**

- Add to list as soon as you get them
- Tell someone before you see them
  - During the day: message in group secure chat
  - After hours: notify senior on call, PGY3/APP/Fellow
- See the consult
- Staff with senior (PGY3/APP/Fellow)
- Staff with attending
- Communicate plan to primary team
- Write your note, consent, post, write orders, f/u on tests

## WEEKLY SCHEDULE

### Monday

**OR:** Jackson

**Clinic:**

MOC 190: PA/NP 9 AM - 12 PM; 1PM-3:30 PM

MOC 190: Ahle 1 PM-4 PM

Fall River: Luks, Renaud and Lorenzo, alternating weeks

### Tuesday

**General Surgery Grand Rounds:** 7-8 AM

**OR:** Renaud

**Clinic:**

MOC 190: Chernoguz 9 AM-12 PM

MOC 190: Monteagudo 9 AM-12 PM and 1-3:30 PM

MOC 190: Luks 1 PM-4 PM

MOC 190: Vascular Anomalies Clinic 12-2 PM, 2<sup>nd</sup> Tues/month

### Wednesday

**General Surgery M&M:** 7-8 AM

**Pediatric Surgery Conference Time:** 7-9:15 AM

2<sup>nd</sup> and 4<sup>th</sup> Wed.; 7-8 AM Pediatric Surgery M&M, 8-9 AM Tumor Board

5<sup>th</sup> Wednesday: Joint conference with Oncology

**OR:** Ahle

**Clinic:**

MOC 190: PA/NP 10 AM – 12 PM

COOP clinic 1<sup>st</sup> floor: Burn Clinic 1 PM – 4 PM

MOC 190: Renaud 1-4 PM

MOC 190: Jackson 9 AM-12 PM; 1-4 PM

### Thursday

**OR:** Monteagudo

**Clinic:**

MOC 190: Renaud 9 AM-12 PM

MOC 190: Ahle 9 AM-12 PM

MOC 190: Chernoguz 1 PM – 4 PM

### Friday

**OR:** Chernoguz

**Clinic:**

MOC 190 Bowel Management 9 AM-12 PM, 1 PM-4 PM

## **ROLE OF THE ADVANCED PRACTICE PROVIDERS (APP)**

The role of the Advanced Practice Provider (APP) on the Pediatric Surgery Service is multi-faceted and incorporates both the inpatient and outpatient environments.

- Outpatient responsibilities include specified clinic days (Monday/Weds/Fri) for post-op visits, wound/ostomy/gastrostomy tube care, bowel management, and new consults at the discretion of the attending.
- Inpatient responsibilities include participation in the care provided by the Pediatric Surgery team.
  - o The APPs collaborate with the entire Pediatric Surgery team to optimize the care of the patient through all phases of hospitalization.

### Roles and responsibilities:

- To round in the NICU and PICU, or Hasbro Floors (depending on coverage), to help as needed, and to support the PGY-1/PGY-3/Fellow
- To see consults as needed both in the PICU/NICU or in Hasbro
- Help formulate and enact a daily plan for all patients seen by the service
- If the Fellow/PGY-3 is unavailable, to act as a senior backup for the PGY-1 seeing floor and ED consults
- To organize the care of all patients going to the OR and to help to ensure their pre-operative preparation is complete (e.g. consent, booking, prep)
- To participate in OR cases and to act as the proceduralist for bedside and sedated procedures as needed
- To respond to all Level A and B trauma activations, and to cover Level C activations as needed by the availability of the junior on the service
- To participate in national organizations in some capacity
- To participate in and ensure active credentialing within specialty/interests (PALS/BLS, ATLS, ABLS, Airway, Bowel management)
- In addition to contributing to the education of rotating residents and medical students, the APP will proctor both PA and nurse practitioner students as scheduling allows

## **PEDIATRIC SURGERY/PICU CO-MANAGEMENT**

Patients in the PICU admitted to the Pediatric Surgery/Pediatric Trauma service (excluding ECMO patients at this time) have collaborative co-management by the Pediatric Critical Care Medicine team.

The co-management process will align with the following guiding principles:

- As stated above, this excludes ECMO patients at this time. However, best practices of communication, collaboration, and sharing of information and tasks remain recommended for ECMO patients in the PICU.
- Patients will continue to have the Pediatric Surgery attending as the attending of record.
- Co-rounding with the Pediatric Surgery and PICU teams is encouraged but often not feasible based on OR schedules. Pediatric Surgery and PICU attendings will touch base on co-managed patients at a minimum each morning.
- Pediatric Surgery and PICU attendings will develop clear expectations for each patient in terms of goals, shared decision-making, and deferred decision-making (such as diet/NPO status decisions deferring to Pediatric Surgery team). For Pediatric trauma patients, all decision-making is deferred to Pediatric Surgery even with co-management as per trauma guidelines.
- Nursing, RT, responding providers (residents and APPs), and support services will be able to contact the PICU team and PICU team will escalate and communicate appropriately with the Pediatric Surgery team. Especially as this process rolls out, erring on the side of more communication is always best.
- Nursing, RT, responding providers (residents and APPs) for the Pediatric Surgery and PICU teams may also always escalate to Pediatric Surgery and PICU attendings for any concerns.
- Decisions regarding intermediate status or suitability for transfer/discharge will remain part of shared decision-making and ultimately is the purview of the attending of record's service.

## TRANSFER OF NICU PATIENTS TO HASBRO

### Elective Transfer for Surgery

When a NICU patient is undergoing a scheduled operation at a future date:

- First, confirm if the patient has an EPIC MRN. Patients may have an account if they have undergone a cardiac echo or had consults to other specialties through the BrownHealth system.
- If the patient does not have an EPIC chart:
- Call NICU at 401-274-4922 and request they fax a demographic sheet to ExpressCare at 401-444-7100 (or fax the sheet yourself).
- Wait at least 5 minutes for the fax to go through, and call ExpressCare at 401-444-3000 to request a chart be created for a future encounter.
- Log into the patient's EPIC chart and click the Prep for Surg/Proc tab (or search via search bar if you can't find it).
- In the Prep for Surg/Proc encounter:
  - Add the Case Request in the Orders section.
  - Add the H&P in the History/Note section.
- Prep the consent including all six attendings and the correct procedure.
- Add a diagnosis at the bottom left corner.
- Sign the encounter at bottom right, adding the attending as cosigner.
- Log into EPIC remotely from W&I and consent the parents electronically in person or by phone.
- If consenting by phone, will require a witness signature.
- Please record the phone number used to contact the parents on the signature portion of the consent.

### Emergent Transfer

If there is an emergent transfer to either PICU or the OR:

- First, confirm if the patient has an EPIC MRN. Patients may have an account if they have undergone a cardiac echo or had consults to other specialties through the BrownHealth system.
- If the patient does not have an EPIC chart:
- Call NICU at 401-274-4922 and request they fax a demographic sheet to ExpressCare at 401-444-7100 (or fax the sheet yourself).
- Wait at least 5 minutes for the fax to go through, and call ExpressCare at 401-444-3000 to request a chart be created for transfer or admission.
- Provide Name, DOB of patient, the diagnosis and name of attending surgeon
- If the patient has an EPIC chart:
- Call ExpressCare at 401-444-3000 to notify about transfer or admission.

- Answer their questions about accepting provider and bed control issues.
- Provide Name, DOB of patient, the diagnosis and name of attending surgeon
- ExpressCare will create an admission via transfer center and start the transfer process.
- For PICU transfer, talk to PICU attending and PICU charge RN to expedite the process.
- For direct to OR transfer, talk to OR board runner at 401-444-5657 to expedite the process.
- Talk to anesthesia board runner to help expedite the process.
- At this point, chart can be accessed and notes, orders, case requests, and consents may be completed as above.

### Non-Emergent Transfer to Hasbro In-Patient Unit

If a surgical NICU patient requires transfer to Hasbro for ongoing care:

- The face sheet from the NICU should be given to the Case Manager at Hasbro at the earliest convenience. They will complete insurance authorization.
- A tentative date of transfer will be set. Non-emergent transfers should be avoided on Mondays and Fridays
- On the day of transfer, Express Care (401-444-3000) needs to be called in order to create the admission account.
- NICU RN to Hasbro RN report should be given via phone or face to face at the time of transfer.
- The NICU discharge summary should be completed by the NICU staff, printed out, and be available to Hasbro staff for review at time of transfer.

## PEDIATRIC SEDATION

### Moderate Sedation

Moderate sedation is a state in which a patient responds purposefully to voice or tactile stimulation and maintains a patent airway and adequate spontaneous ventilation. This is further defined as the administration of 2 agents in combination to achieve a desired effect, i.e. administering a narcotic in conjunction with a benzodiazepine.

- Moderate sedation for procedures may **NOT** be provided on the general care units.
- The provider of moderate sedation cannot concurrently perform the planned procedure unless a separate credentialed professional (RN or LIP) is entirely dedicated to monitoring the patient at all times.
- The proceduralist does not need to have credentials for moderate sedation when the sedation is being provided and monitored by a credentialed individual.

### Analgesia:

For patients that require wound care or other scheduled care that is painful, narcotics can be considered and administered. Depending on the patient's status IV or PO can be used; this should be discussed with a senior or attending prior to ordering. Medications commonly used on the general floors:

- IV morphine (0.05mg/kg-0.1mg/kg)
- PO oxycodone liquid or tablet (0.1mg/kg, 2.5mg x1 Dose, 5mg x1 dose)

Dilaudid can be considered, but use of this medication must be discussed with senior team members/attending prior to ordering/administering.

Fentanyl is NOT approved for the general floors.

### Anxiolysis

For patients that require care that could be stressful or anxiety inducing (i.e. placement of nasogastric tube), a benzo can be considered. Again, discuss with your senior or attending before use. On the general floors:

- IV Versed (0.05mg/kg- 0.1mg/kg)
- Intranasal Versed (if unable to get IV access) (0.2mg/kg-0.3mg/kg)
- IV Ativan (0.05mg/kg- 0.1mg/kg)
- PO Ativan (0.05mg/kg- 0.1mg/kg)

For any escalation of care to include moderate sedation or deep sedation, the Pediatric Sedation Team or Pediatric Anesthesia team should be notified. Consultation of these teams requires prior discussion with a Pediatric Surgery senior members including the appropriate Pediatric Surgery Attending.

These patients would need to be NPO:

- Clear Liquids: 2 hours
- Breastmilk: 4 hours
- Infant Formula: 6 hours
- Nonhuman milk: 6 hours

-Regular diet: 8 hours

Pediatric Sedation: ext 4-6091

Under the direction of the Pediatric Critical Care service, this team can provide moderate and deep sedation so that the patient will spontaneously breathe and not require intubation. Agents used by this team include but are not limited to fentanyl, propofol, ketamine, versed and precedex. The patient must have IV access prior to arrival in the Pediatric Sedation suite; if Anesthesia is providing sedation, an IV is required prior to arrival in the PACU. A consultation order for Pediatric Sedation should be placed through EPIC, but this must be followed by a call to Pediatric Sedation by an APP, the fellow, or the attending.

## COMMONLY USED MEDICATIONS

### Emergency Medications

Adenosine (3 mg/ml)	0.1 mg/kg (total max single dose 6 mg)
Atropine (0.4 mg/ml)	0.02 mg/kg (min 0.10 mg; max 0.5 mg)
Calcium Chloride 10% (100 mg/ml)	20 mg/kg
Epinephrine-Bradycardia (1:10,000)	0.1 cc/kg via IV, IO
Epinephrine-Bradycardia (1:1000)	0.1 cc/kg via ETT
Glucose/Insulin (8U regular insulin)	1 ml/kg bolus in 50 ml 50% glucose)
Lidocaine 1% (10 mg/ml)	1 mg/kg bolus IO/IV
Infant NaHCO <sub>3</sub> 4.2% (0.5 mEq/ml)	1mEq/kg
Narcan (1 mg/ml)	0.1 mg/kg
Amiodarone	5 mg/kg bolus IV/IO (repeat up to 2x for refractory VF/pulseless VT)
Procainamide	15 mg/kg over 30-60 min IV/IO
KCl	0.5 mEq/kg

### ICU Medications

Versed	0.1 mg/kg Drip: 0.05-0.1 mg/kg/hr
Fentanyl	1 mcg/kg Drip: 0.5-1 mcg/kg/hr
Morphine	0.1 mg/kg Drip: 0.05-0.1 mg/kg/hour
Vecuronium	0.1 mg/kg Drip: 0.1 mg/kg/hr
Precedex	Drip: 0.025 mcg/kg/hr titrate to effect
Milrinone	Drip: 0.25-0.75 mcg/kg/min
Dopamine	Drip: 2.5-20 mcg/kg/min
Dobutamine	Drip: initial dose 0.5-1 mcg/kg/min then 2-20 mcg/kg/min
Epinephrine 1:1000	Drip: 0.05-1.0 mcg/kg/min
Nipride	Drip: 0.5-8 mcg/kg/min
Norepinephrine	Drip: 0.1-1.0 mcg/kg/min

## General Medications

Acetaminophen	10-15mg/kg q6hr IV/PO/PR
Ibuprofen	10mg/kg q6hr PO
Toradol (Ketorolac)	0.5mg/kg/dose q6hr IV; max 15mg, max 5 days
Protonix ( $\geq 15$ kg to $< 40$ kg)	0.5mg/kg q12hr IV Over 40 kg: 40 mg/dose
Zofran	0.15mg/kg IV max 16mg/dose
Morphine	0.1mg/kg q3hr IV
Oxycodone	0.15mg/kg/dose q4-6hr PO
Augmentin (dosed on Amoxicillin)	20-40mg/kg/day divided q6-8hr PO
Zosyn	80mg/kg/dose q6-8hr IV
Vancomycin	15mg/kg q6-12hr IV
Bactrim (dosed on trimethoprim)	5mg/kg/day q12hr PO max 40 mg
Clindamycin	10mg/kg TID IV/PO
Pepcid	0.5mg/kg/day PO

## Post-op pain control

### Baseline starting regimen:

Acetaminophen and an NSAID may be alternated every 3 hours to help provide more continuous coverage:

PO to be used unless NPO and/or NGT to suction

Acetaminophen (15 mg/kg up to 650 mg) PO/PR/IV every 6 hours; optimally scheduled at 05:00-11:00-17:00-23:00 or 00:00-06:00-12:00-18:00

If not NPO: Motrin (10 mg/kg up to 400 mg) PO every 6 hours; if NPO: Toradol (0.5 mg/kg up to 15 mg) IV every 6 hours; optimally scheduled at 02:00-08:00-14:00-20:00 or 03:00-09:00-15:00-21:00 depending on acetaminophen dosing. DO NOT give to children < 6 months old

### Adjuncts

Lidocaine patch (1-2) scheduled daily: apply to sides of incisions, not above or below

Gabapentin (100-300 mg) or Lyrica (25-50mg) scheduled TID

Oxycodone (2.5-5mg) every 6 hours PRN for severe pain only

Morphine (weight-based dosing) every 4 hours PRN for severe pain only

# ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA

Alejandra M. Casar Berazaluce, MD, MSc, FAAP

## Introduction

Surgical management of esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is one of the greatest successes in pediatric surgery of the last century. This diagnosis went from a 100% mortality rate prior to 1940 to a >90% survival rate today (>98% in BW>1.5kg without cardiac anomalies).

## Epidemiology

- EA +/- TEF occurs in approximately one in 3500 live births.
- 40% of these patients are born prematurely, presumably secondary to polyhydramnios resulting from the inability to swallow amniotic fluid.
- Mean gestational age is 36.5 weeks, and mean birth weight is 2.5 kg.
- 40-60% will have associated anomalies (incl. VACTERL spectrum).

## Embryology

EA/TEF is caused by a complex and poorly understood process that likely involves environmental, biomechanical, and genetic factors resulting in abnormal foregut development. During the fourth week of gestation, the ventral portion of the foregut gives rise to the trachea and lungs while the esophagus develops from its dorsal aspect. Failure of the process of foregut septation leads to TEF.

## Pathophysiology

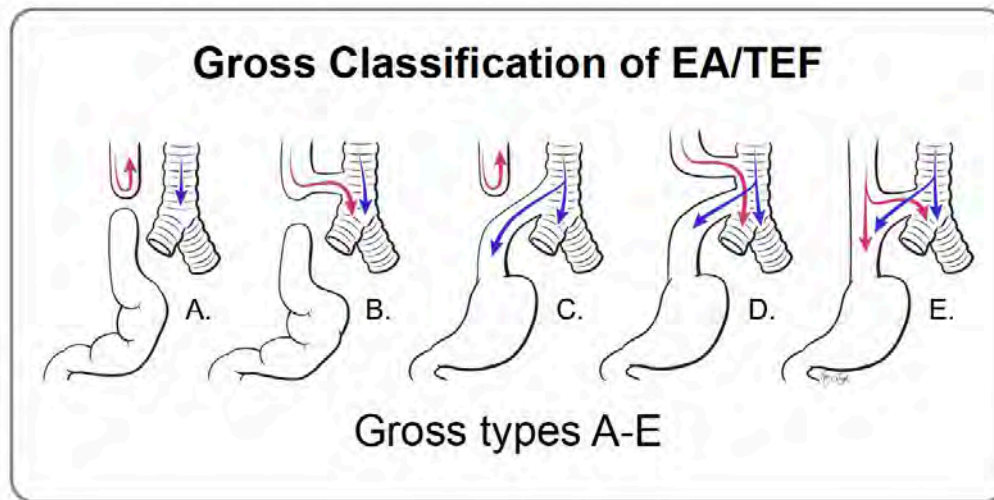
Uncorrected EA/TEF can lead to a variety of sequelae including inability to feed and handle secretions, pneumonitis/pneumonia, worsening respiratory mechanics, and cardiac arrest secondary to aspiration and abdominal distention -> Death.

## Classification

The most common variant is a proximal EA with distal TEF (Type C). *Long gap EA* is poorly defined and has been qualified as greater than two vertebral bodies, greater than 3 centimeters, or inability to perform a primary repair at birth.

### *Gross's Anatomical Classification*

<i>Type</i>	<i>Description</i>	<i>Incidence</i>
A	Esophageal atresia <i>without</i> tracheoesophageal fistula	8%
B	Esophageal atresia <i>with proximal</i> tracheoesophageal fistula	<1%
C	Esophageal atresia <i>with distal</i> tracheoesophageal fistula	87%
D	Esophageal atresia <i>with proximal and distal</i> fistula	1%
E	Tracheoesophageal fistula <i>without atresia</i>	4%



Type E is also known as “H-type fistula”: shaped more like an N because opening is proximal in the trachea and distal in the esophagus, leading to late diagnosis (usually recurrent pneumonia especially with reflux). It can be assessed via bronchoscopy and esophagoscopy or prone pull-back esophagogram.

## Presentation

Most cases of esophageal atresia and tracheoesophageal fistula present in the newborn period with excessive drooling and feeding difficulties.

- Inability to pass an NG/OG tube.
- Secretions and feeds lead to coughing, choking, cyanosis, tachypnea.
- Positive pressure ventilation may lead to abdominal distension (except in cases without distal fistula, where the abdomen will be scaphoid).

## Assessment

A complete assessment of the newborn with suspected esophageal atresia and tracheoesophageal fistula includes an evaluation for associated chromosomal and anatomic anomalies related to the VACTERL spectrum.

- Complete history and physical
- Replogle placement by experienced provider (usually stops around 10cm)
- Babygram to assess Replogle tip position and presence of gas in the abdomen

## Preoperative Workup

The goal of the initial evaluation is to identify all the serious anomalies and rank them in order of severity and priority of treatment. Physical exam, plain XRs, and echocardiogram are mandatory.

The most important part of the evaluation is an echocardiogram due to a ~50% incidence of cardiac anomalies with increased mortality.

- This should also assess the side and character of the aortic arch and look for vascular anomalies, such as interrupted IVC.
- If arch or vascular anomalies are identified on echo, 3-D imaging is recommended with CTA or MRA.

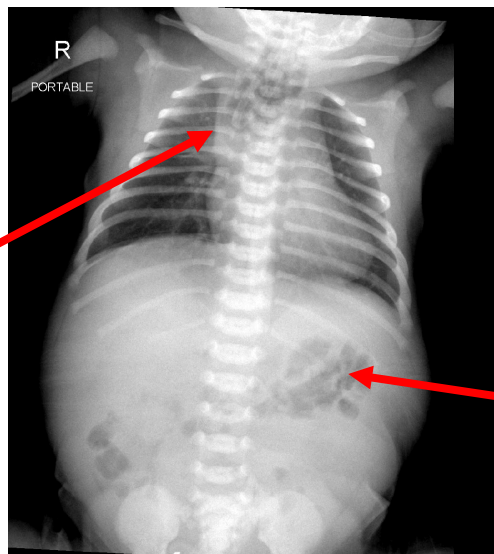
Complete VACTEGRL workup includes:

- **Vertebral** (tethered spinal cord, bony anomalies)
  - Physical exam
  - Spine ultrasound before 3 months (MRI after 3 months)
  - Spine and sacral x-rays
- **Anorectal** (imperforate anus)
  - Physical exam
- **Cardiac** (ASD, VSD, other structural anomalies)
  - Echocardiogram
- **TracheoEsophageal Fistula**
  - Plain films after NG/OG placement
- **Gynecology/Genitourinary**
  - Physical exam
  - Pelvic ultrasound at thelarche
- **Renal** (hydronephrosis, duplicate/absent kidneys)
  - Renal ultrasound
- **Limb Anomalies** (radial anomalies)
  - Physical exam
  - Plan x-rays

## VACTERL-G Screening

- Physical Exam (V, A, L, G)
- Thorax/abdomen X-ray with feeding tube (TE)
- AP/Lateral sacral X-ray (V)
- Ultrasound (V, R, G)
  - Spine < 3months (MRI after 3 months)
  - Renal
  - Pelvic @ Thelarche
- Echocardiogram (C)

Coiled NG =  
Esophageal atresia



Air in GI tract =  
Tracheoesophageal  
fistula

## Prenatal Diagnosis

An absent or small gastric bubble in combination with polyhydramnios is suspicious for EA/TEF. Polyhydramnios is typically not seen until after 24 weeks gestation. These findings have low sensitivity and specificity, with a PPV ~50%.

Identification of a dilated proximal esophageal pouch or a distended hypopharynx on US are a more reliable finding, better confirmed by MRI after 26wks gestational age, with a sensitivity of ~95%. Despite advances in US technology, only 20-30% of cases are diagnosed before delivery.

Because of the association with other anomalies, amniocentesis, fetal echocardiogram, and fetal MRI are recommended for any suspected fetal cases.

There are currently no standard fetal interventions, but prenatal diagnosis is an opportunity to optimize perinatal management including delivery at a tertiary center.

## Medical Management

The goal is to preserve circulation and oxygenation prior to repair. Routine positive pressure ventilation should be avoided to avoid gastric distension.

- Strict NPO
- Nasal or oral esophageal sump tube to suction
- Elevate HOB 30 degrees to prevent reflux and aspiration
- Initiate gastric acid suppression
- Avoid intubation and provide supplemental oxygen
- If intubation is required due to extreme prematurity or cardiac anomalies, it should be a multidisciplinary discussion – avoiding positive pressure, potentially intubating past the fistula, or considering alternatives like HFJVL
- If higher pressures are utilized and severe gastric distension occurs, it may lead to inability to ventilate, which may require emergent bedside gastric decompression
- Create a low stimulation environment to prevent crying and aerophagia
- Judicious fluid resuscitation
- PICC and TPN as there may be a delay to enteral feeding

## Surgical Management

Neonates with TEF and critical ventilation challenges should be considered for emergent fistula division +/- gastrostomy placement. Outside of this, VACTERL workup should be completed and associated anomalies should be considered when determining the timing of the repair. Multidisciplinary discussions and subspecialty consultations should be obtained.

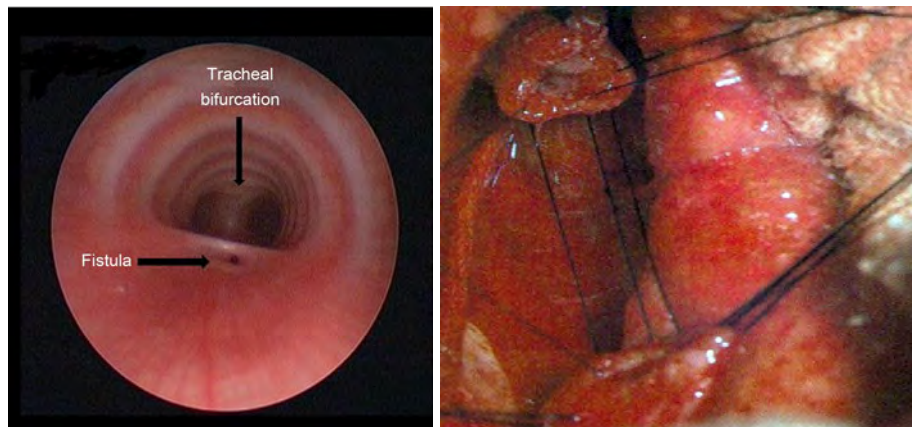
Patients with a gasless abdomen are unlikely to have a distal TEF, and long gap EA should be suspected. Because growth of both segments continues during the first few months of life, delayed primary repair is an option, and a gastrostomy tube should be placed with the goal of reaching goal bolus feeds.

Neonates under 1-1.5kg should also be managed with gastrostomy and delayed repair.

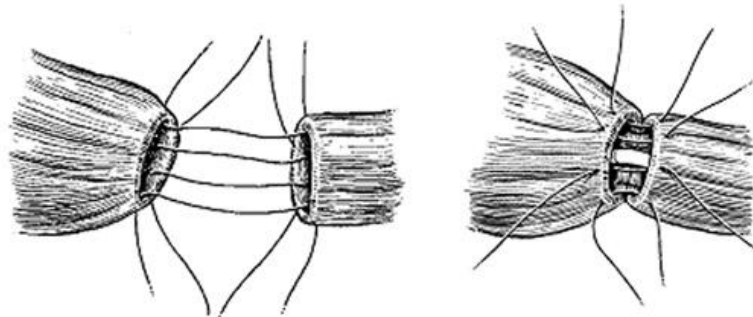
## Operative Intervention

- Direct laryngoscopy and rigid bronchoscopy can be performed to assess for laryngeal, tracheal, and bronchial anomalies and laryngo- and tracheomalacia in addition to evaluation for proximal and distal TEFs.
- Localization of the TEF on endoscopy determines the optimal placement of the endotracheal tube during intubation, as well as placement of a balloon catheter to block the fistula.
- Flexible esophagoscopy and gastroscopy are not routine but can be used as adjuncts to define anatomy in complex cases and when intraoperative concerns arise.
- Standard positioning is left lateral with a thick axillary roll and padding of all pressure points. The arm is extended above the head or prepped into the field.
- Sterile prep should include from the ear superiorly to the umbilicus inferiorly and from left of the spinal processes posteriorly to left of midline anteriorly.
- A right posterolateral thoracotomy is performed, usually at the fourth intercostal space.
- Muscle sparing techniques are available to preserve the latissimus dorsi.
- An extrapleural approach is common. After division of the intercostal muscles, the pleura is pushed away with moist gauze or cotton tipped applicators to access the posterior mediastinum.
- Self-retaining retractors are placed to maintain exposure ensuring no undue pressure is applied on critical structures.
- Control of the fistula:
- The distal esophagus is localized anterior to the spine and below the azygos vein, to the left or slightly anterior to the aorta, between the left and right vagus nerves.
- The TEF is usually behind the azygos vein. Although dividing the vein can help with exposure, it is best to avoid it, and it can be an important source of venous drainage in complex anatomy
- A vessel loop is placed around the TEF.
- Esophageal mobilization:
- The upper pouch is identified anterior to the spine in the apex of the chest.
- The SVC, right subclavian, vagus and phrenic nerves should be identified and protected.
- A catheter/instrument/endoscope can be placed transorally to help with identification and dissection.
- A traction suture is placed at the apex for mobilization.
- The distal esophagus is mobilized up to the TEF on the tracheal side, being cautious of a 1-2cm common wall by staying close to the esophagus.
- Aggressive mobilization was previously discouraged, but mobilizing down to the diaphragm is now considered safe and reduces tension at the anastomosis.
- The phrenoesophageal ligament should not be disrupted as it leads to hiatal hernia and severe GERD.
- Tracheoesophageal fistula repair:
- Fine, absorbable sutures are placed transversally across the TEF.
- The fistula is sequentially divided in small bites and closed, leaving a 1 mm cuff.
- A leak test is performed with Valsalva at 30-40 cm H<sub>2</sub>O pressures.

- Esophageal anastomosis:
- The lower esophagus is trimmed to good tissue and a reasonable lumen.
- A primary single-layer end-to-end anastomosis is performed with interrupted fine absorbable suture.
- Corner sutures are placed first for alignment.
- The posterior row of sutures is placed, then crossed to distribute tension before tying.
- The anterior row of sutures is then placed, crossed, and tied.
- Ensure the mucosa does not extend outside the anastomosis.
- A chest tube may be placed depending on surgeon preference. Their utility is controversial. Transanastomotic feeding tubes are now discouraged.
- The chest wall is closed in layers.
- A thoracoscopic MIS approach is available but not yet standard. Dissection is transpleural in this case.



*L: Fistula on preoperative rigid bronchoscopy; R: Esophageal anastomosis*



### For proximal TEF:

- The ET tube should be placed below the fistula.
- The tracheal repair is transverse.
- The esophageal repair is longitudinal.
- Ensuring the suture lines don't overlap may reduce recurrence.
- One option is to rotate the esophageal repair 90 degrees.
- A posterior tracheopexy (suturing the membranous trachea to the anterior longitudinal ligament of the spine) may allow separation of the suture lines and aid in correction of tracheomalacia.

### For isolated TEF:

- The repair is done with a right transverse neck incision.
- Dissection of the cervical esophagus is medial to the carotid sheath.
- A tissue flap may be interposed after suture repair to prevent recurrence.

### For insufficient esophageal length:

- Flexion of the neck or spine may help facilitate the anastomosis.
- Circular myotomies can be performed in the thickened upper esophageal pouch.
- A tubularized anterior flap can also be created from a dilated upper pouch.
- Gastroplasties can be performed to bring some stomach into the chest, but a high risk of intractable reflux makes this less favorable.
- Several traction techniques have been explored.
- Internal traction can be performed by suturing both ends together without creating the anastomosis or suturing them to the spine to allow for longitudinal growth under tension, with anastomosis 4-6 weeks later.
- External traction can be performed via the Foker process, externalizing the sutures through the chest wall to serially increase the traction over weeks.
- Esophageal replacement techniques are utilized when the native esophagus cannot be preserved.

### Postoperative Care

- Most babies return to NICU intubated and are supported with TPN and broad-spectrum antibiotics. The use of neuromuscular blockade is uncommon.
- Judicious extubation – do not extubate until cleared by surgeon
- Avoid CPAP, NIPPV, and HFNC in the setting of fresh tracheal repair and esophageal anastomosis
- Increases risk of leak and mediastinitis
- Avoid traumatic reintubation in the setting of postoperative edema and a fresh anastomosis
- Continue gastric acid suppression and HOB elevation 30-45 degrees
- Gentle oropharyngeal suctioning to clear accumulation of secretions due to anastomotic edema – not too deep as it may hurt the anastomosis
- Chest tube in place – to water seal or suction per operating surgeon, to stay in place until after oral feeds are initiated
- Strict NPO and TPN
- Broad-spectrum antibiotics – duration per operating surgeon
- Trans anastomotic tubes are debatable, but may be used for gastric decompression and/or post-anastomotic enteral feeding; they must be carefully secured and must not be replaced if dislodged
- Pain control with IV or PR Tylenol; minimize narcotics to prevent ileus and prolonged mechanical ventilation
- Consider contrast esophagogram between POD 5-10 to evaluate anastomosis before initiation of oral feeds

### Complications

- The most important complications are anastomotic leak and stricture.

- Anastomotic leak rate is around 20%
- Most are contained and not clinically significant
- If not contained, may lead to sepsis
- Higher incidence in high tension anastomosis or significant dissection causing devascularization and ischemia, such as with long gap atresia
- May be diagnosed with routine esophagogram or finding of frothy saliva from chest tube – fluid amylase will be elevated
- Most can be managed nonoperatively with IV antibiotics, antifungals, TPN, and chest tube drainage
- Repeat esophagogram is usually obtained weekly, and most leaks will close without intervention in 2-4 weeks
- Anastomotic stricture rate is around 40%
- Dilation as early as 2-4 weeks after repair is the first line therapy
- Balloon catheter dilation (8-10mm diameter) under fluoroscopy
- Will likely need repeat dilations
- Need GERD management, may not improve if untreated
- Persistent strictures can be managed with intralesional steroids or mitomycin C
- Recurrent TEFs occur in around 5% of cases
- Usually present with pneumonia or respiratory issues associated with feeds
- Diagnosis can be made by prone pull-back esophagogram or bronchoscopy
- Require operative intervention
- Vocal cord dysfunction occurs in around 3% and can be uni- or bilateral
- Preoperative bronchoscopy establishes baseline function
- Injury is more common after reoperation
- Can present with cyanotic spells, with a differential diagnosis of recurrent aspiration or tracheomalacia

## Outcomes

In the absence of major cardiac anomalies and low birth weight, EA/TEF survival is high. Unfortunately, long term morbidity remains the norm among survivors.

### Risk Stratification

<i>Class</i>	<i>Risk</i>	<i>Clinical Characteristics</i>	<i>Mortality</i>
I	Low	BW >2kg	0%
II	Intermediate	BW >2kg + cardiac anomaly	7%
III	High	BW 1-2kg + cardiac anomaly <i>or</i> BW <1kg	33.3%
IV	Super High	BW <1kg + cardiac anomaly	100%

Long term problems:

- Pulmonary issues: reactive airway disease, bronchitis, pneumonia
- Gastrointestinal issues: Dysphagia, chronic GERD, dysmotility
- Scoliosis (secondary to thoracotomy, not particular to EA/TEF)
- Long-term follow up includes GERD management and screening for Barrett's esophagus.

### Long Gap Esophageal Atresia

 <p>Fluoro 2wks post G-tube</p>  <p>No length consensus</p> <p><b>DIAGNOSIS</b></p>	 <p>Delayed Primary Repair (Weeks-Months)</p> <p>If unsuccessful,</p>  <p>Elongation    Substitution</p> <p><b>MANAGEMENT</b></p>	 <p>↑ Esophagitis ↑ Barrett's ↑ Cancer</p>  <p>EGD every 10yrs</p>  <p>Monitor: dysphagia, respiratory function, growth &amp; nutrition</p> <p><b>FOLLOW UP</b></p>
 <p>Journal of Pediatric Surgery</p>	<p>Baird, R., Lal, D. R., Rizzo, R. L., Diefenbach, K. A., Downard, C. D., Shelton, J., ... Goldin, A. (2019). Management of long gap esophageal atresia: A systematic review and evidence-based guidelines from the APSA Outcomes and Evidence Based Practice Committee. <i>Journal of Pediatric Surgery</i>, 54(4), 675-687. <a href="https://doi.org/10.1016/j.jpedsurg.2018.12.019">https://doi.org/10.1016/j.jpedsurg.2018.12.019</a></p>  <p>@alejandracaser</p>	

# NEC (NECROTIZING ENTEROCOLITIS)

## Background/Terminology/Epidemiology

Suspecting and recognizing NEC in the NICU is prudent while you are on your pediatric surgery rotation as it is both the most common newborn surgical emergency and the most common surgical cause of death in the NICU population with a mortality rate of approximately 25%. It is a disease of prematurity and has an incidence of about 1/1000 live births.

In addition to prematurity, risk factors include absent or reversed end diastolic umbilical artery blood flow, maternal eclampsia, fetal distress, premature rupture of membranes, physiologic derangements of the newborn (asphyxia, hypothermia, respiratory distress, apnea), congenital heart disease, persistent fetal circulation, PDA, sepsis, gastroschisis and Hirschsprung disease.

The development of NEC is not completely understood and is multifactorial. It results from mucosal compromise in the setting of pathogenic bacteria. This most often occurs following the introduction of enteral feeds.

Human breast milk reduces the relative risk of NEC as compared to formula feeds. Probiotic use has been shown to reduce the incidence of NEC in very low birth weight infants; however, the lack of FDA regulation of probiotics makes them hard to use in the hospital setting.

Stage	Systemic Findings	Abdominal Findings	Radiographs
IA (Suspected)	Perinatal stress, temperature instability, apnea, bradycardia, lethargy	Feeding intolerance, distension, occult blood in stool	Normal, intestinal dilation, mild ileus
IB (Suspected)	Same as above	Gross blood in stool	Same as above
IIA (definite, mildly ill)	Same as above	Absent bowel sounds, tenderness	Pneumatosis intestinalis
IIB (definite, moderately ill)	Metabolic acidosis, thrombocytopenia	Cellulitis, mass	Same as above plus ascites
IIIA (advanced, severely ill, intact bowel)	Hypotension, bradycardia, respiratory/metabolic acidosis, DIC, neutropenia	Same as above plus peritonitis	Same as above
IIIB (advanced, severely ill, perforated bowel)	Same as above	Same as above	Pneumoperitoneum

Necrotizing Enterocolitis. (2021). In Hirschl, R., Powell, D., & Waldhausen, J. (Eds.), *Pediatric Surgery NaT*. American Pediatric Surgical Association. [https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829043/all/Necrotizing\\_Enterocolitis](https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829043/all/Necrotizing_Enterocolitis)

## Diagnostic Work up

Presentation: Neonates with NEC present with signs and symptoms of abdominal sepsis. They often have intolerance to feeds, including emesis, in the setting of abdominal distension and tenderness. They sometimes have abdominal wall discoloration including ecchymosis, erythema or edema. As they progress,

they can develop peritonitis and physiologic derangements such as hypotension requiring vasopressors and increasing respiratory support.

Imaging: First line imaging is a babygram, which is an XR on the baby's body. Signs of NEC include pneumatosis (air in the wall of the intestine) and/or portal venous gas (air in the portal vein.) This imaging gets serially followed, often every 6-8hrs, until it stabilizes or improves. An additional finding may be pneumoperitoneum which would prompt urgent peritoneal drain placement or laparotomy. Occasionally an abdominal US will be obtained. This is not the gold standard as it has not been validated that pneumatosis seen on US but not on XR is clinically significant.

Labs: Neonates consume platelets at a faster rate than they can create them when sick; because of that, thrombocytopenia occurs. Low or dropping platelet count is an important indicator of disease progression in NEC. You will additionally follow your normal labs for acidosis such as lactate and a blood gas.

This all comes together to create the modified Bell Staging Criteria help categorize patients with NEC (table).

## Treatment

If the neonate does not have an acute abdomen (hemodynamic instability requiring vasopressors, abdominal wall ecchymosis/erythema, peritonitis, pneumoperitoneum on XR) we begin with non-operative management. This will apply to ~75% of patients:

- NPO
- Replogle to suction for gastric decompression
- Broad spectrum antibiotics (Zosyn)

If the neonate has pneumoperitoneum we consider bedside peritoneal drain placement vs exploratory laparotomy based on their age, weight and how they are doing overall. Final decision is based on clinical judgement, however, most neonates <1kg will be offered drain placement.

If the neonate has an acute abdomen or is failing non operative management, an exploratory laparotomy is required. These babies are often managed in stages. At the first operation, frankly necrotic bowel is excised with marginal bowel left in situ and the abdomen left open. This process is repeated until all bowel has declared itself. Often times, the final operation involves creation of one or multiple ostomies and a mucous fistula at the time of abdominal wall closure with a goal of future stomal reversal. In the acute phase, the goal is to preserve as much bowel as possible.

Sometimes at exploration, we find *NEC totalis*, in which >80% of the bowel is ischemic. This finding is not compatible with life. No further intervention is able to be offered and a sensitive, compassionate conversation with the family is required to discuss end-of-life care.

## Post op monitoring

In the acute phase, the goal is to stabilize the neonate: wean vasopressor support, wean ventilator support and recover bowel function.

Up to 10% of neonates can develop recurrent NEC

In the intermediate phase, we focus on allowing the neonate to get enteral nutrition and grow. This can be challenging depending on where in the intestine the NEC occurred and the location of the ostomy; those neonates with short gut or proximal stomas will require long term TPN and may require feeding with

refeeding of the mucous fistula. A reasonable goal for stoma reversal is 6-8 weeks following abdominal closure and when the infant has reached 2kg.

Infants with NEC are at risk for stricture formation. The data is mixed, but a stricture can form in up to ~60% of neonates who had NEC. The risk is highest in those infants managed medically. Strictures are a result of scarring of prior ischemic bowel as it heals. It presents as worsening PO intolerance as feeding volume increases. For those neonates with stomas, contrast studies (barium enema and small bowel follow through) of the intestine is considered prior to reversal to rule out stricture formation.

## Long term Complications

In neonates that require extensive resection, they are at risk for intestinal failure and short bowel syndrome. This requires multidisciplinary care with nutrition and gastroenterology. It often requires gastric feed supplementation with a gastrostomy tube and parenteral nutrition with a central line.

## Resources and Suggested Readings

Pediatric Surgery NaT chapter on NEC:

Necrotizing Enterocolitis. (2021). In Hirschl, R., Powell, D., & Waldhausen, J. (Eds.), *Pediatric Surgery NaT*. American Pediatric Surgical Association. [https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829043/all/Necrotizing\\_Enterocolitis](https://www.pedsurglibrary.com/apsa/view/Pediatric-Surgery-NaT/829043/all/Necrotizing_Enterocolitis)

Holcomb and Ashcraft's Pediatric Surgery:

Fisher, Jeremy G., & Moss, R. Lawrence. (2020). 33 - Necrotizing Enterocolitis. In *Holcomb and Ashcraft's Pediatric Surgery* (Seventh Edition, pp. 536–556). Elsevier Inc. <https://doi.org/10.1016/B978-0-323-54940-0.00033-X>

# MECONIUM ILEUS

## Terminology

### Simple meconium ileus:

Extensive plugging of the intestinal tract, including small and large bowel, with thick, inspissated, tar-like meconium

### Complicated meconium ileus:

Pre- or perinatal presentation of meconium ileus complicated by meconium peritonitis, volvulus and/or intestinal atresia

### Cystic fibrosis:

95% of cases of true meconium ileus occur in patients with cystic fibrosis. Conversely, 20% of infants with cystic fibrosis present with meconium ileus. (A syndrome that is identical in presentation to meconium ileus can occur without associated cystic fibrosis – most often in patients of Eastern-Asian descent.)

### Meconium ileus equivalent:

Obstipation syndrome in older children with cystic fibrosis, in whom thick stools can cause an acute intestinal obstruction

### Meconium plug syndrome:

A completely different pathology, characterized by *normal* meconium that is blocked from evacuation by a thick, whiteish luminal plug (often in mid- or proximal colon). Once the plug is evacuated, stooling is normal. Meconium plug is NOT associated with cystic fibrosis. It is sometimes used interchangeably with Small left colon syndrome. Both these conditions can be associated with Hirschsprung disease (10%).

## Diagnosis

### Prenatal history and diagnosis:

- Familial history of cystic fibrosis in 30% (both parents carriers, but not themselves affected; male patients are sterile)
- Prenatal diagnosis may be based on the following, in addition to genetic testing for CF: intestinal dilation, echogenic bowel, calcifications and pseudocyst (the latter two in case of complicated meconium ileus).

### Presentation, evaluation, and treatment:

#### Babies with meconium ileus present with abdominal distension

- As this is a congenital condition, meconium ileus presents at birth
- Like most forms of lower intestinal obstructions, the main feature is severe abdominal distension, sometimes associated with (bilious) vomiting. (In upper intestinal causes of obstruction in

newborns such as malrotation, abdominal distension is less prominent, and the abdomen can even be scaphoid.)

- Differential diagnosis includes anorectal malformations (imperforate anus), Hirschsprung disease, ileal and colonic atresia and midgut volvulus.
- On palpation, the abdomen feels doughy (“putty sign”) – unlike distal intestinal atresia or anorectal obstructions, where the abdomen is tympanic. In complicated meconium ileus, there may be significant discoloration and edema of the abdominal wall, from irritation by (prenatal ) meconium peritonitis (figure below).



## Imaging:

Note: IV fluid resuscitation is crucial before any contrast study

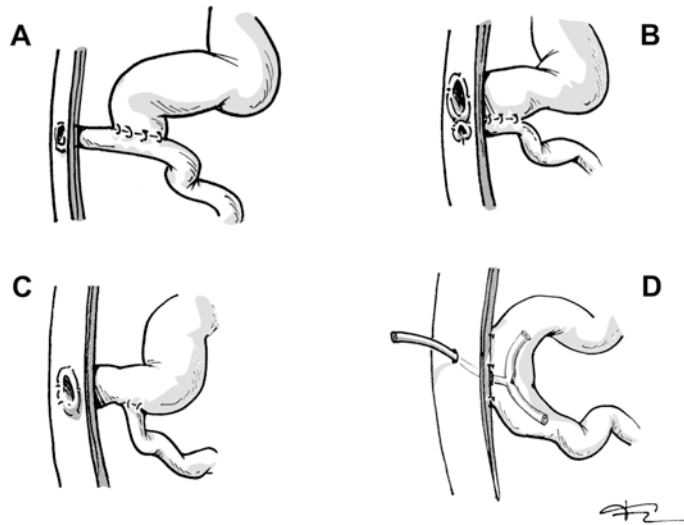
- KUB
- Multiple gas-distended intestinal loops (as with other forms of lower GI obstructions)
- Superimposed “soap bubble” or “ground glass” opacities representing meconium-filled loops
- Absence of air-fluid levels on cross-table lateral, as the air is trapped in the tenacious meconium and does not rise to the top
- Speckled calcifications or calcified rim of a pseudocyst, if complicated meconium ileus is present
- Ultrasound
- Abdominal ultrasound is not necessary, but can add to the diagnosis: calcifications (from prenatal perforation) may be more obvious by U/S
- Contrast enema
- If meconium ileus is suspected, a rectal contrast study is indicated
- **NO barium should be used** (in case of intestinal perforation, barium peritonitis will severely complicate the condition and treatment)
- An iso-osmolar water-soluble contrast enema will show a microcolon (distal to the obstruction) with filling defects caused by pellets of stool (figure above).

## Treatment

- Non-operative management is indicated in a stable newborn **without signs of complicated meconium ileus**.
- Hyperosmolar water-soluble contrast enema can be successful in dislodging the thick meconium through its hygroscopic properties, drawing fluid into the intestinal lumen

- **Adequate IV fluid resuscitation is crucial, as the osmotic effect of the enema can rapidly cause cardiovascular collapse in the borderline-dehydrated infant:** at least 150 mL/kg/d in a full-term neonate (if already euvolemic); follow with urine output and heart rate (urinary catheter mandatory); oro- or nasogastric decompression (Replogle), IV antibiotics, Type & Crossmatch for 2 U of PRBC.
- Successful therapeutic enema **requires** reflux of contrast into the meconium-filled dilated small bowel loops.
- Dislodgement of a (whiteish) meconium plug, followed by copious evacuation of normal-appearing meconium, suggests meconium plug syndrome. No further treatment may be necessary – but 10% of these patients may prove to have Hirschsprung disease
- Operative treatment is indicated in any of the following situations:
  - Complex meconium ileus with evidence of (prenatal) intestinal perforation
  - Intestinal perforation during non-operative attempts (contrast enema)
  - Inability to reflux contrast into the dilated small bowel loops (suspect atresia)
  - Inability to resolve the intestinal obstruction despite several enema attempts

## Operative maneuvers



- Laparotomy: typically, transverse supra-umbilical incision – offers widest exposure
- Clearing the obstructing meconium
- Enterotomy of the (distal) ileum and manual evacuation of the (mostly sterile) thick, tarry meconium – in a cephalad and caudad direction. Can be a tedious and lengthy maneuver; avoid serosal or mesenteric trauma
- Irrigation of the intestine with warm saline, with or without added acetylcysteine (Mucomyst®), 4 mL of a 10% solution, diluted to 10 mL with normal saline
- Primary closure of the enterotomy if entire bowel is effectively cleared of meconium
- **However**, most surgeons will prefer to leave access to the intestines for post-operative

irrigation:

- Bishop-Koop (proximal end-to-distal side, or “distal chimney”) enterostomy (A)
- Divided (double barrel) ostomy (B)
- Santulli (proximal side-to-distal end, or “proximal chimney”) enterostomy (C)
- T-tube (D)
- Factors to consider in choosing a stoma technique:
- Evacuation of tenacious meconium is easier through a large enterotomy or transected bowel – leading to either a double-barrel or chimney stoma
- If all meconium is evacuated, a T-tube may be best
- In case of atresia and diameter discrepancy between proximal and distal bowel, a Bishop-Koop enterostomy allows primary repair with a “pop-off” and irrigation valve
- Appendectomy is not mandatory, but often performed: it can be an elegant way to an irrigation enterostomy, and pathology exam can help in the diagnosis
- Intestinal resection
- In case of frankly ischemic bowel (volvulus) or atresia, resection is necessary.
- Bowel resection should be limited, if possible, to avoid short gut syndrome (consider second-look for borderline ischemic loops)
- Anastomosis: see above – primary anastomosis without decompression stoma is rarely safe
- Meconium peritonitis
- Prenatal perforation often leads to a meconium pseudocyst
- Overly aggressive lysis of dense adhesions may jeopardize bowel length
- Proximal stoma (and mucous fistula) may be the safest first-line treatment

## Postoperative care

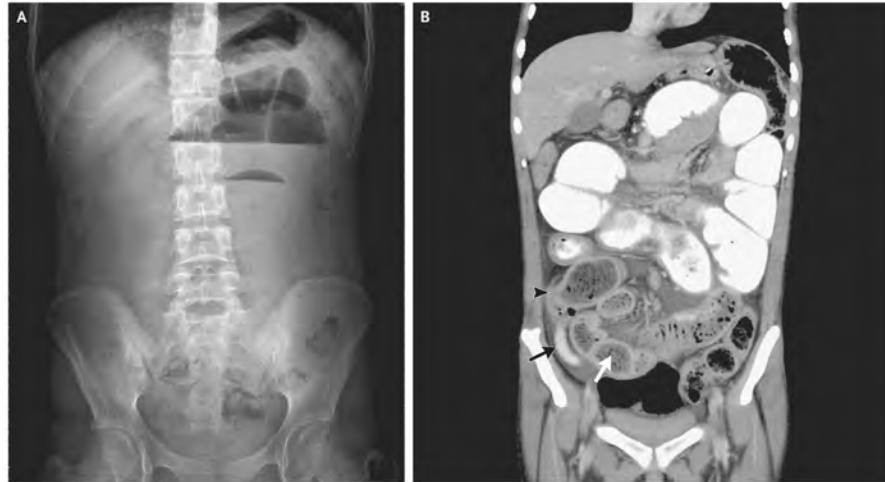
### Immediate postoperative period

- Continue Iv antibiotics for 48 h, or longer if clinically indicated
- Adequate postoperative hydration: Depletion of the intravascular space is bound to continue as water is drawn into the bowel
- Start irrigations via distal stoma or NG tube with 2-4% Mucomyst® on 3<sup>rd</sup>-4<sup>th</sup> postoperative day – daily dose, until full feeds
- Early consultation with pulmonary/CF specialist
- Confirmation of CF diagnosis (sweat test at 1 month, typically)
- Transition to enzyme replacement once tolerating feeds

### Long-term management

- Infants with CF who present with meconium ileus are NOT at greater risk of severe disease – in fact, neonatal presentation (because of meconium ileus) may allow early preventive measures to avoid recurrent pulmonary complications
- Meconium ileus equivalent is a form of intestinal obstruction in older children and adults with

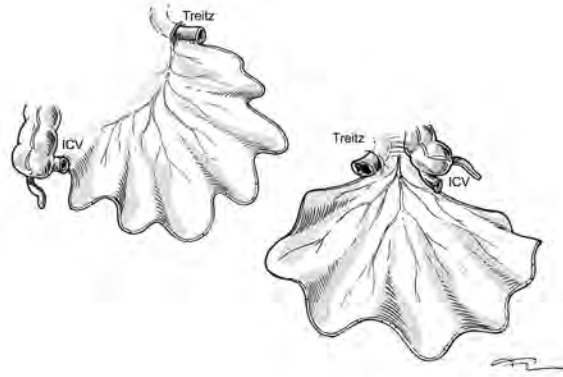
CF, mimicking the neonatal condition (figure below). Hyper-osmolar enema treatment can be attempted, but aggressive IV fluid hydration must precede the procedure, as cardiovascular collapse is a significant risk.



### Suggested Readings:

1. Kronfi R, Maguire K, Walker GM. Neonatal stomas: does a separate incision avoid complications and a full laparotomy at closure? *Pediatr Surg Int* 29: 29:299–303, 2013.
2. Carlyle BE, Borowitz DS, Glick PL. A review of the pathophysiology and management of fetuses and neonates with meconium ileus for the pediatric surgeon. *J Pediatr Surg* 47:772-81, 2012.
3. Burke MS, Ragi JM, Karamanoukian HL, et al. New strategies in nonoperative management of meconium ileus. *J Pediatr Surg* 2002;37:760-764.
4. Fakhoury K, Durie PR, Levison H, et al. *Arch Dis of childhood* 1992;67:1204-1206

## MALROTATION AND MIDGUT VOLVULUS



### Terminology

#### Key features of normal intestinal rotation:

- 1) the duodenum describes a “C-loop” with the third portion of the duodenum (at the ligament of Treitz) to the left of the midline,
- 2) the superior mesenteric artery runs in front of the third portion of the duodenum,
- 3) the mesentery is attached posteriorly along a broad line that runs from the ligament of Treitz (left upper quadrant) to the cecum (right lower quadrant), and
- 4) the colon describes a frame with the cecum and ascending colon fixed along the right side of the abdomen and the descending colon fixed along the left side.

#### Malrotation:

- most commonly used to describe non-rotation: failure of the duodenum to form its characteristic “C-loop” and instead coursing in a straight cephalocaudal line into the proximal jejunum.
- Other rotational anomalies: incomplete rotation, reverse rotation, and errors of intestinal fixation, typically of the cecum and the ascending and descending colon.
- Incidence of isolated malrotation: 1 in 500 live births; much more common in a number of genetic, chromosomal and congenital disorders.
- **Errors of intestinal rotation usually do not affect intestinal function**
- **They do place a child at risk of midgut volvulus – a life-threatening that causes acute obstruction of the root of the SMA, resulting in ischemia of the entire midgut from the ligament of Treitz to the ascending colon.**
- **Midgut volvulus demands rapid operative correction!**

### Diagnosis

#### Presentation:

##### Bilious emesis

- Suspect midgut volvulus in **ANY** infant or newborn presenting with bilious vomiting.

- Seen in 1/2 of the infants presenting with malrotation or midgut volvulus, and in 1/3 of those older than one month.
- Not pathognomonic for malrotation: seen with duodenal atresia, annular pancreas, small bowel atresia, meconium ileus, Hirschsprung disease

#### Physical exam/other findings

- Scaphoid abdomen
- May be absent later in the course due to gastric distention, distention of ischemic bowel, ascites
- Bloody stools (late findings)
- Tachycardia, poor capillary refill
- Oligo or anuria
- Metabolic acidosis on labs

#### Imaging:

##### KUB

- Non-specific
- Can suggest presence of malrotation/volvulus due to “double bubble” from tight Ladd’s bands, although distal gas can be present
- NEVER used alone to rule out volvulus; if suspicion high, go to UGI

##### Upper gastrointestinal contrast series (UGI)

- Most reliable diagnostic method
- Done on stable patients; unstable patients with peritonitis should go to the OR
- Normal rotation: duodenal C-loop crosses the midline, ligament of Treitz (duodenojejunal junction) located to the left of the spine and at least as high as the pylorus
- Malrotation or non-rotation: ascending and transverse colon are located to the left of the spine; also, right-sided position of the majority of small bowel loops and absence of typical colonic frame
- Midgut volvulus: corkscrew (or apple peel) appearance of the first jejunal loops, best seen in a lateral view

##### Ultrasound



- **Not** the gold-standard for detecting malro or volvulus
- Has become more accurate over time
- Identification of 3<sup>rd</sup> portion of duodenum crossing spine suggest normal rotation

- Superior mesenteric artery (SMA) posterior and to left of vein in normal rotation
- Malro: SMA to right of SMV
- Midgut volvulus: torsion of vessels cine as a swirl or “whirlpool” pattern on Doppler ultrasound

#### Contrast enema:

- Less reliable: partially or completely rotated colon does not rule out duodenal malrotation

#### CT:

- Can see whirlpool sign and duodenal obstruction
- Not the diagnostic procedure of choice, especially for neonates

### Treatment

Immediate intervention for **midgut volvulus** diagnosed by UGI or strongly suspected (acutely obtunded infant with bilious vomiting)

- IV fluid resuscitation,
- NGT
- foley
- Do the preceding as you **GET THE PATIENT TO THE OR**
- Urgent intervention for malrotation without obstruction in otherwise well patient
- UGI may miss intermittent volvulus or partial obstruction

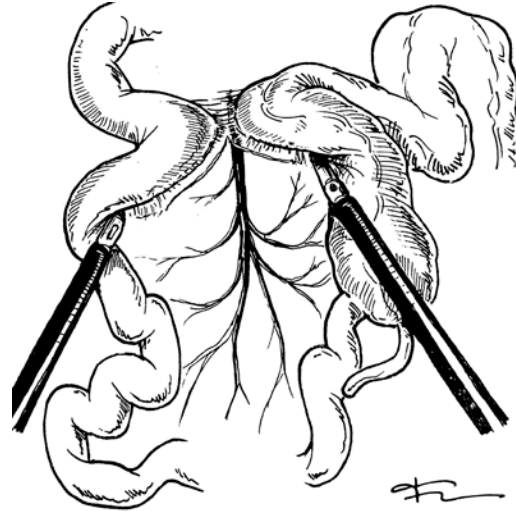
### Surgical repair

Midgut volvulus should be treated with *open* repair; *laparoscopy* can be considered for elective or semi-urgent operation for malrotation.

#### *Open repair*

- Generous transverse upper abdominal incision for newborns and infants, midline laparotomy for older children
- Intestinal contents are exteriorized completely and assessed.
- If volvulus is present, the small bowel must be rotated in a **counterclockwise** fashion to undo (“Turn back the clock”)
  - Even if initially ischemic, detorsion and decompression can produce significant or complete return of normal color and perfusion.
  - Detorsion is complete when the base of the mesentery is visualized (small bowel will be right of midline and colon left)
- Ladd’s bands: may run from the cecum to the right abdominal wall. Can obstruct the duodenum or duodenojejunal junction. If present, they are divided
- Avascular attachments between the duodenum and the ascending colon are divided. This allows the duodenum to fall back in the right gutter and the ascending colon to the left.
- Operation is complete when the main trunks of the SMA and SMV are exposed (figure right).
- In the stable patient, appendectomy is performed as well
- If **bowel necrosis** is present, resection may be necessary. Guiding principles:
- If no contamination and limited extent of necrosis, resection with primary anastomosis is safe

- If spillage or significant contamination or the patient is unstable, ostomies are safer
- In case of extensive ischemia, limited resection, if possible, followed by second-look might preserve some bowel length



## Suggested Readings

- Ladd WE. Congenital obstruction of the duodenum in children. *N Engl J Med* (1932) 206:277-283
- Kluth D, Kaestner M, Tibboel D, Lambrecht W. Rotation of the gut: fact or fantasy? *J Pediatr Surg* (1995) 30:448-453
- van den Brink GR. Hedgehog signaling in development and homeostasis of the gastrointestinal tract. *Physiol Rev Surg Int* (2007) 87:1343-1375
- Seashore JH, Touloukian RJ. Midgut volvulus. An ever-present threat. *Arch Pediatr Adolesc Med* (1994) 148:43-46
- Patino MO, Munden MM. Utility of the sonographic whirlpool sign in diagnosing midgut volvulus in patients with atypical clinical presentations. *J Ultrasound Med* (2004) 23:397-401
- Spigland N, Brandt ML, Yazbeck S. Malrotation presenting beyond the neonatal period. *J Pediatr Surg* (1990) 25:1139-1142
- Lessin MS, Luks FI. Laparoscopic appendectomy and duodenocolonic dissociation (LADD) procedure for malrotation. *Pediatr Surg Int* (1998) 13:184-185

## ANORECTAL MALFORMATIONS

A range of defects in which there is a lack of anal opening, most often with an associated rectal fistula to various points along the perineum or GU tract.

- Incidence 1/3000-1/5000
- Male:female ratio 3:1

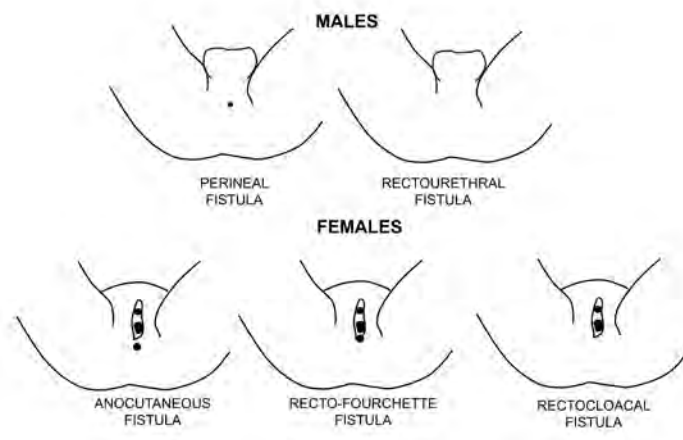
Most common: male – rectourethral fistula; female- rectovestibular fistula

Without fistula: 5%, associated with Trisomy 21

### Types:

**Male:** rectoperineal fistula, rectourethral fistula (bulbar, prostatic), recto-bladder neck fistula

**Female:** rectoperineal fistula, rectovestibular (-fourchette) fistula, cloaca



### Associated abnormalities:

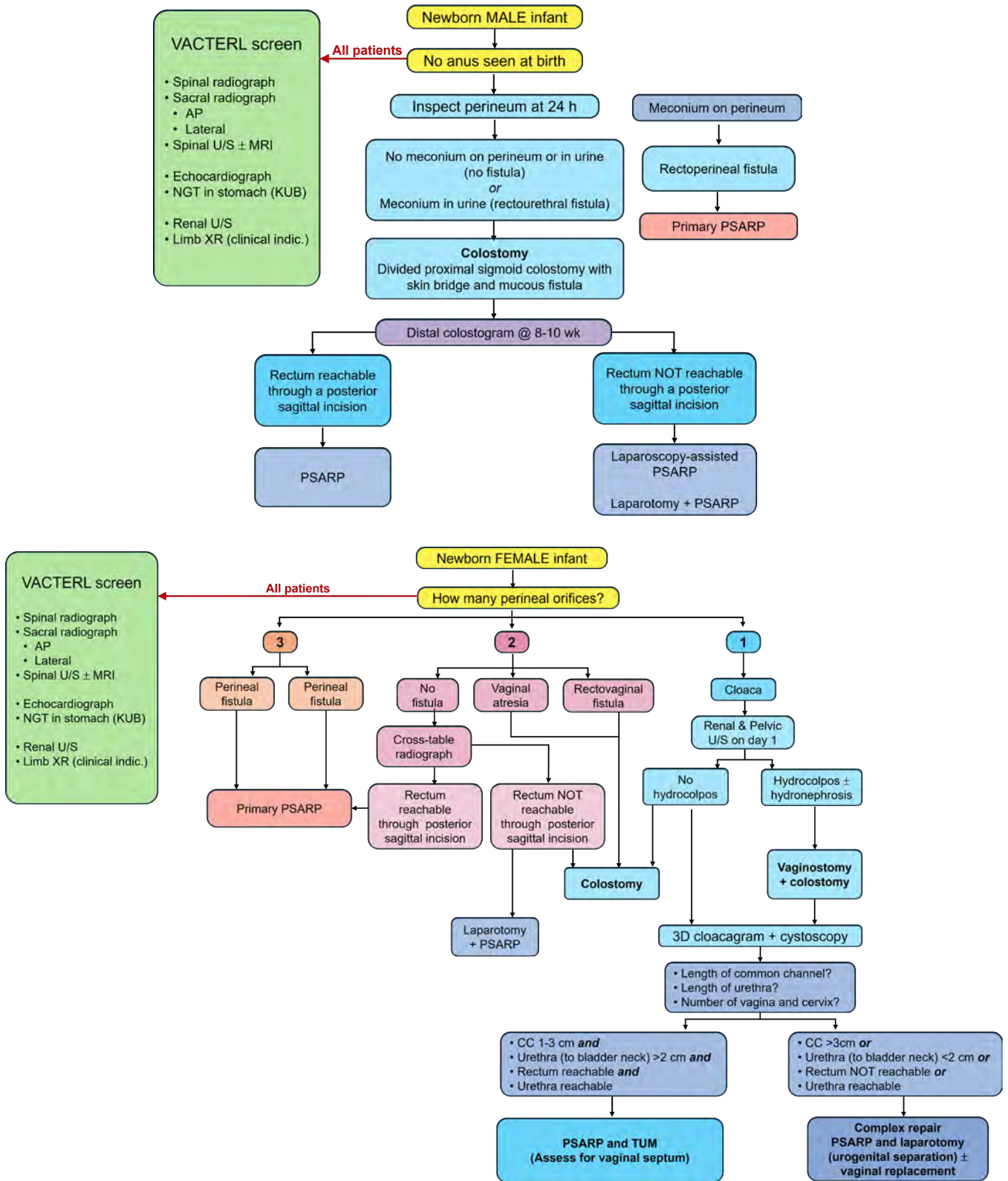
Most common: urological (increased likelihood with higher malformations)

Also: gynecological (female, particularly vaginal septum); cardiac; gastrointestinal (esophageal atresia, duodenal atresia); spinal; skeletal

### Preop workup

Goals are to classify the anomaly, recognize and manage life-threatening anomalies, and determine operative management.

- Physical exam
- Place NG tube (rule out esophageal atresia)
- ECHO
- Renal ultrasound
- Spinal x ray (AP/Lateral sacral x ray)
- Abdominal ultrasound (female – evaluate for hydrocolpos)
- Spinal ultrasound



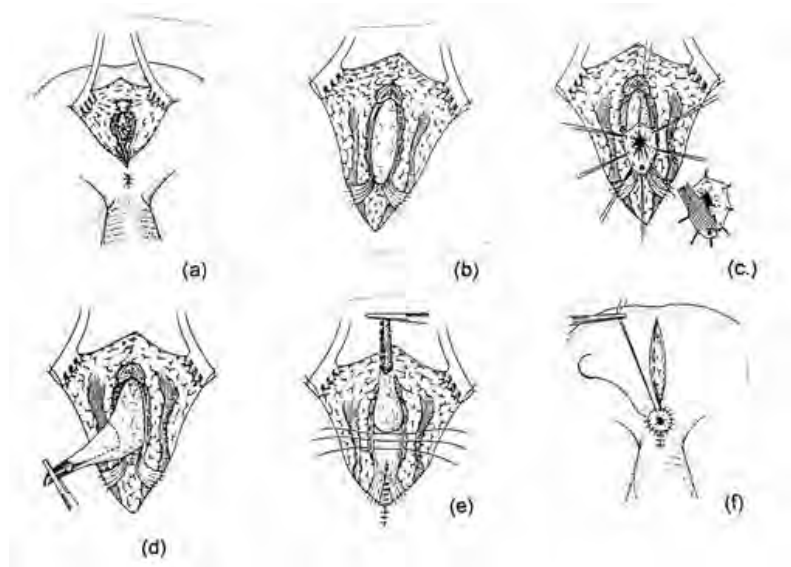
From Rentea R, Smith C, Chan S, Zahora P. Anorectal malformation basics. 2024 Annual PCPLC Meeting, November 2024.

## Management

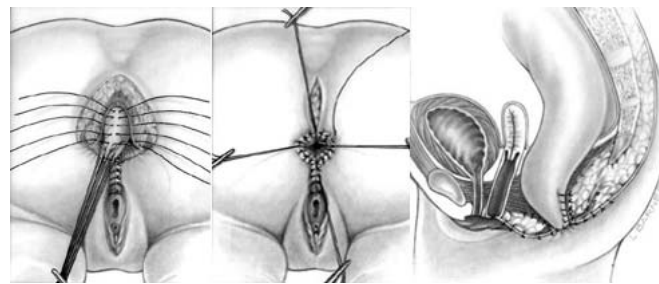
Perineal fistula, vestibular fistula: anoplasty, limited posterior sagittal anorectoplasty (PSARP)

Vestibular fistula: Primary PSARP or initial colostomy

Rectourethral fistula, rectobladder neck fistula, cloaca: initial colostomy, later PSARP and other needed reconstruction



*Male PSARP*



*Female PSARP – rectovestibular fistula*

### Potential surgical complications:

- Urethral injuries
- Vas deferens injury
- Dehiscence
- Posterior urethral diverticulum
- Recurrent epididymitis and orchitis
- Constipation
- Incontinence
- Prolapse

# PYLORIC STENOSIS

## EPIDEMIOLOGY

Hypertrophic pyloric stenosis occurs in 1 in 250 children. It is usually not present at birth, but typically develops in the first few weeks of life. There appears to be a slight familial incidence, although no clear pattern of inheritance is seen, and sporadic cases are more common than familial ones. There is a strong male preponderance, and first-born infants tend to be more often affected. If pyloric stenosis occurs in a girl, her children are at a significantly increased risk of contracting the condition. Epidemiologic studies have failed to identify environmental, genetic or dietary (breast milk vs. formula) factors. Pyloric stenosis is more common in full-term infants; if preterm infants are affected, they are typically several weeks older than their term counterparts.

## DIAGNOSIS

Pyloric stenosis appears to be a progressive disease. It is now well documented that the full-blown syndrome, in which pyloric obstruction is complete, is often preceded by a period of intermittent pyloric spasm. The infant first vomits feeds (but not water or Pedialyte<sup>®</sup>), and this may be interpreted as intolerance to a particular type of formula. Not uncommonly, formula changes will have been tried before pyloric stenosis is suspected. Increased awareness among pediatricians means that pyloric stenosis is now diagnosed several weeks earlier, on average, than 20 years ago.

### History and presentation

- Hallmark of pyloric stenosis is projectile, non-bilious vomiting
- Mean age at onset is 2 to 3 weeks of life, but vomiting may be intermittent for another 1-2 weeks
- Absence of bile helps rule out other, more distal, causes of intestinal obstruction, such as midgut volvulus or Hirschsprung's disease.
- Coffee ground vomiting suggests associated gastritis and is often a later finding.
- Non-surgical conditions such as sepsis, gastroenteritis, hepatitis are easily eliminated because the child with pyloric stenosis appears otherwise well and extremely hungry
- Not associated with diarrhea or abdominal tenderness
- Mild jaundice is common (secondary to glucuronyl transferase deficiency)
- In progressive disease, dehydration, lethargy and shock ensue

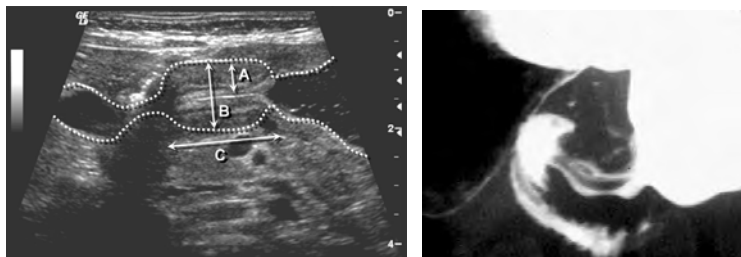
### Physical exam

- Palpation of the "olive" (hypertrophic pylorus) *should* be possible in all cases, but requires patience and a cooperative infant
- Examiner should stand on the patient's left; it helps to distinguish the right kidney (gentle, deep palpation) and the rectus muscles, to differentiate them from the pylorus. The pylorus is gently teased from under the liver edge, and rolled between the fingers and the infant's spine
- Having the child in a parent's lap, in a quiet environment and calmed with pacifier or a few sips of clear fluid can be useful

- If the stomach is full, one-time nasogastric aspiration can be helpful – after the infant is calmed
- In advanced cases, strong gastric peristalsis can be visible in the left upper quadrant (rare sign)

## Laboratory and imaging

- Urinalysis confirms mild or moderate dehydration and, in advanced cases, aciduria
- CBC is helpful, since 3-4-week-olds have physiologic anemia (transition from fetal to adult Hb)
- Serum electrolyte panel is most important: hypokalemia, hyponatremia, alkalosis and dehydration can be confirmed and corrected (atraumatic blood drawing (i.e, no heel stick) will avoid hemolysis and erroneously elevated serum K



- Ultrasound is typically performed even before surgery is consulted (by PMD or ED). Confirmatory findings include A) pyloric muscle thickness  $>3$  mm, B) total pyloric diameter  $>14$  mm and C) length of pyloric channel  $>16$  mm: mnemonic 3.1416 ( $\pi$ ) – although cut-off values can vary by institution (figure 1). Strong gastric peristaltic waves and a pyloric channel that never opens are additional findings
- UGI is almost never needed, but shows the historically classic features: mushroom sign, “inverted 3” or umbrella sign and railroad track sign (figure 2)

## TREATMENT

### Preoperative management

- o Pyloric stenosis is **not** an operative emergency
- o Correction of fluid and electrolyte anomalies is mandatory before surgery – even if it takes 1 or more days
- o Severe cases (dehydration, shock, severe electrolyte anomalies, paradoxical aciduria) require PICU admission
- o Nasogastric decompression is not routinely necessary
- o Intravenous hydration with 0.45 N saline, or 0.9 N in severe cases
- o Supplementation with KCl (10-30 mEq/L) after urine output is documented
- o Correct any electrolyte anomaly gradually

### Pyloromyotomy

- o Even when NPO, an infant with pyloric stenosis is considered to have a full stomach

- Gastric aspiration immediately before rapid induction (or, less commonly, awake intubation with cricoid pressure) is safest
- Perioperative antibiotics are not necessary
- Regardless of the approach (open or laparoscopic) the principle is the same: extramucosal splitting of the pyloric muscle over its entire length

Open pyloromyotomy:

- Curved incision in the upper umbilical crease. A little cheating (slightly wider than the natural crease) allows a sufficiently long incision
- The subcutaneous tissues are dissected in the supra-umbilical midline (taking care to go immediately superficial to the linea alba). The fascia is opened longitudinally (midline), and can be extended around the umbilicus (2-3 cm long incision)
- If the stomach is not easily seen, the corner of a dry sponge is introduced in the abdomen: omentum will stick to the sponge, and gently pulling on the omentum will deliver the transverse colon, followed by the stomach
- The stomach is grasped along the greater curvature (stomach should be decompressed) and “run” toward the pylorus. This can be done with fingers (with or without an opened, wet sponge for better grip) or with instruments: Singli or Tuttle
- As more distal stomach is exteriorized, its proximal portion needs to be pushed back in the abdomen
- Gentle rocking of the antrum, with or without retraction of the right corner of the wound, will deliver the pylorus



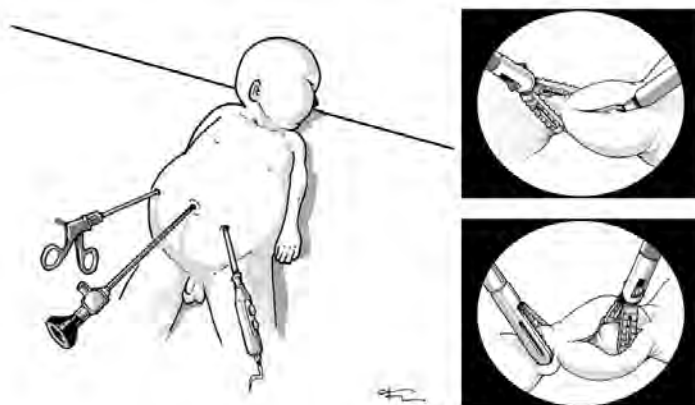
- This maneuver can be frustrating, and the incision may have to be increased; but too large an incision makes it hard to keep the pylorus out of the abdomen
- Once delivered, the pylorus is immobilized with the non-dominant index finger and thumb on either side of the pyloric channel (figure 3)
- A *very shallow* incision is made over the entire length of the pylorus, anteriorly, from stomach to the vein of Mayo at the junction with the duodenum.
- One arm of an opened mosquito clamp, with tips turned up, is used to deepen the pyloromyotomy **by pulling, never pushing** the clamp within the serosal incision. Next, the pyloric spreader (or one arm of it) is inserted inside the pyloric incision, and the spreader is rotated 90 degrees to lie perpendicular to the pyloric channel: this widens the incision enough to allow the spreader to be placed deep into the pyloric incision as in figure 3. The spreader is

then gently, but persistently spread to force the muscle fibers to split and the mucosa to “pop out” into the pyloric wall incision.

- Care must be taken not to spread too forcefully and deeply at either end of the pyloric channel, where gastric and duodenal mucosa, respectively, form a more superficial recess that could be injured
- Completeness of the pyloromyotomy is confirmed by the ability to move each half of the pyloric muscle independently
- Gastric content (fluid and/or air, administered through a nasogastric tube if necessary) is squeezed through the pylorus and into the duodenum, to search for mucosal leaks
- Should a mucosal tear occur, it is repaired with an absorbable suture (4-0 or 5-0 PDS or similar). Omentum can be placed over the pylorus in Graham patch fashion. In the unlikely event that the repair compromises the results of the pyloromyotomy, the pylorus can be rotated 180° along its axis, exposing its infero-posterior surface, and a second pyloromyotomy can be performed
- Some bleeding may be encountered; it is almost always worsened by venous congestion (from the exteriorized pylorus), and stops once the pylorus is returned to the abdomen. Hemostasis, if necessary, should be done carefully
- The fascia is closed with an absorbable suture like 2-0 vicryl. If the linea alba appears weak or flimsy superiorly, a stay suture to delineate the upper aspect of the fascial incision may be helpful
- Skin is closed with a subcuticular suture or dermabond

#### Laparoscopic pyloromyotomy:

- The infant may be positioned perpendicularly on the operating table
- An intraumbilical incision is made to accommodate a 4 or 5 mm diameter, 30 degree telescope and its cannula
- Two additional incisions are made, in each upper quadrant, for 3 or 5 mm diameter instruments. Some surgeons forgo accessory cannulas and insert the instruments directly through the abdominal stab wounds
- The abdomen is insufflated at a maximum pressure of 8-10

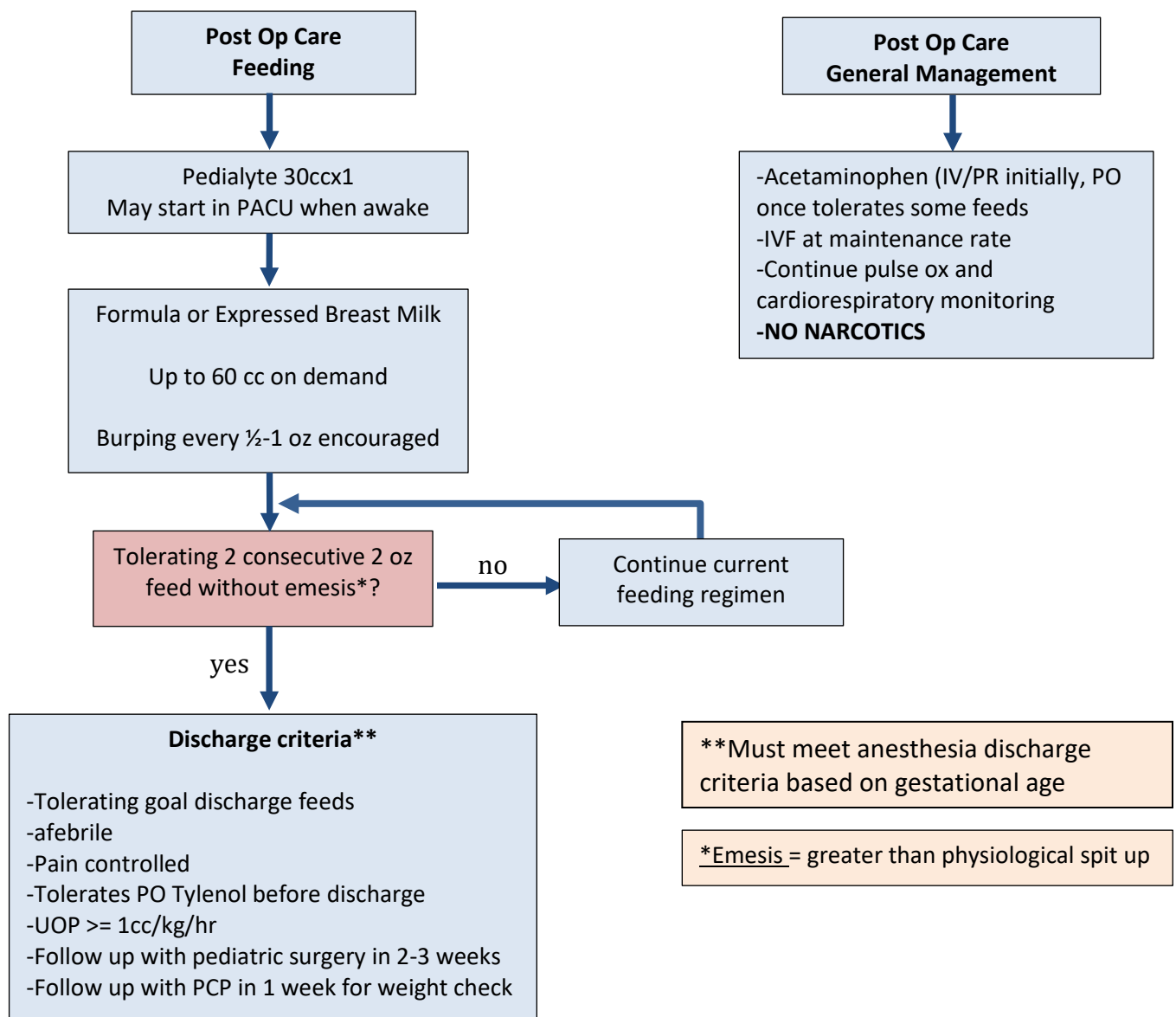


- Once the pylorus is visualized, it is incised longitudinally with a laparoscopic or arthroscopic knife.

- A laparoscopic pyloromyotomy clamp (with serrations on the outside) is then used to spread the muscle fibers as with the open technique, while a grasper stabilizes the pylorus (figure 4)
- At the end, air is insufflated through a naso- or orogastric tube to check for mucosal leaks

## POSTOPERATIVE CARE

- A nasogastric tube is not necessary postoperatively
- Please see following **Post op care and discharge criteria** for postop guideline
- Some vomiting is still expected postoperatively, either from gastric spasms if the child has been ill for a prolonged period of time or, more commonly, because of coexisting gastro-esophageal reflux



## COMPLICATIONS

- Wound infection and fascial dehiscence: should be rare, but seems more common than would be expected with a clean wound (2-3% and as high as 5-10% in some series)
- Unrecognized mucosal perforation and peritonitis are very serious complications, that must be avoided by meticulous technique and intraoperative leak test
- Recurrent pyloric stenosis almost never happens – but may be seen if postoperative feeds (not just Pedialyte ®) were withheld for too long

## Suggested Reading

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*Three historic papers on the first description of the condition and its operative treatment*
4. Vanderwinden JM, Mailleux P, Schiffmann SN, et al. Nitric oxide synthase activity in infantile hypertrophic pyloric stenosis. N Engl J Med 1992;327:511-5.
5. Chen EA, Luks FI, Gilchrist BF, et al. Pyloric stenosis in the age of ultrasonography: fading skills, better patients? J Pediatr Surg 1996;31:829-30.
6. Teele RL, Smith EH. Ultrasound in the diagnosis of idiopathic hypertrophic pyloric stenosis. N Engl J Med 1977;296:1149-50.  
*The first description of ultrasonography for pyloric stenosis, establishing this modality as the new gold standard in pyloric imaging*
7. Kim SS, Lau ST, Lee SL, et al. The learning curve associated with laparoscopic pyloromyotomy. J Laparoendosc Adv Surg Tech A. 2005;15:474-7.
10. Kim SS, Lau ST, Lee SL, et al. Pyloromyotomy: a comparison of laparoscopic, circumumbilical, and right upper quadrant operative techniques. J Am Coll Surg. 2005;201:66-70.
11. Hall NJ, Van Der Zee J, Tan HL, et al. Meta-analysis of laparoscopic versus open pyloromyotomy. Ann Surg. 2004;240:774-8.
12. Hulka F, Harrison MW, Campbell TJ, et al. Complications of pyloromyotomy for infantile hypertrophic pyloric stenosis. Am J Surg 1997;173:450-2.

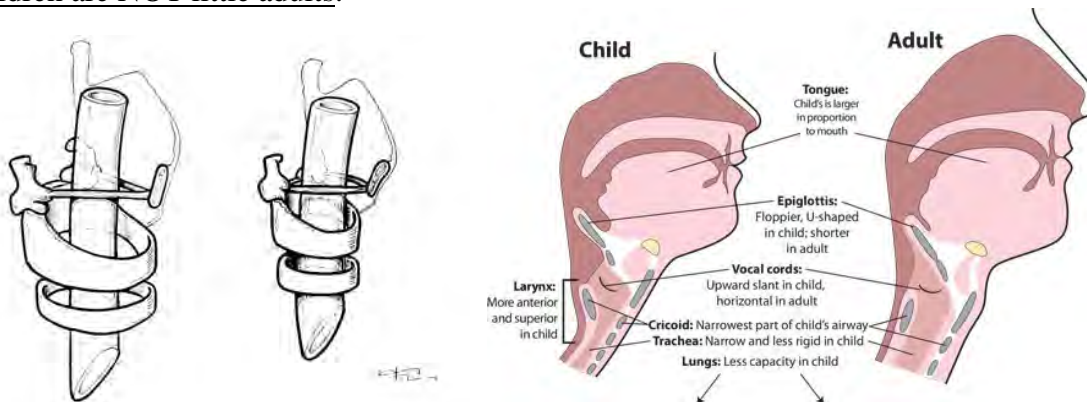
## PEDIATRIC VENTILATORY MANAGEMENT

Signs of Airway Obstruction	
poor movement of air	tracheal tug
faint or absent breath sounds	intercostal retractions (between ribs)
substernal retractions (below sternum)	subcostal retractions (below rib cage)
use of accessory muscles	rocking in chest /abdominal motion
stridor	lack of chest rise
head bobbing	nasal flaring
cyanosis	lack of end-tidal CO <sub>2</sub>

**Indications:** high respiratory rate, labored breathing decreased level of consciousness, inability to protect airway, hypercarbia, chest trauma, aspiration

**Airway Obstruction:** Infants and toddlers tire easily when they have airway or respiratory compromise. Respiratory distress can easily progress to respiratory failure

**Children are NOT little adults:**



- Children have smaller, shorter airways.
- Increased airway resistance → increased work of breathing.
- Pediatric airway is anterior / superior. Glottis opening higher (C2/C3 vs C6 adult)
- Children have larger tongues relative to oropharynx. Posterior displacement → obstruction
- Epiglottis is long, narrow, floppy, and angled away from axis of trachea
- Narrowest portion in children is below vocal cords at level of cricoid cartilage and larynx is funnel shaped. Narrowest portion for adults is glottis inlet and larynx is cylindrical

### Ventilator Variables:

- **Trigger:** ventilator-triggered or patient-triggered breath
  - o The variable that causes inspiratory valve to open (start of breath).
  - o Patient-assisted: Patient makes an effort to take a breath and ventilator senses and supports this (senses negative inspiratory pressure / or inspiratory flow)
  - o Machine-controlled: Ventilator decides frequency of breaths based on set respiratory rate.

- **Limit:** flow (volume) limited or pressure-limited (Fig. 1)
- Flow/Volume-limited: A fixed tidal volume / flow rate is given and maintained throughout inspiration to ensure adequate tidal volume
- Pressure-limited: Pressure is not allowed to surpass a set limit
- **Cycling:** volume, time, flow, pressure cycled
- Volume, time, flow and pressure stop inspiration / start expiration

## Types of Ventilation

### Assist Control:

- Ventilator senses patient breaths and delivers set volume or pressure at set rate in addition to patient-triggered breaths.
- Mandatory breaths: Vent delivers preset volume / flow rate at a set rate. In addition to this, the vent will deliver the preset volume / flow for every additional breath the patient takes. Ex: Rate set at rate 18, Vt 80; if patient breathes 28 breaths / min, the additional 10 breaths would receive a Vt of 80.
- Pressure Control: Volume delivery varies, Inspiratory pressure constant, Inspiratory flow varies. Good for neonates / infants <6-8kg.
- Volume Control: Volume delivery constant; Inspiratory pressure varies; Inspiratory flow constant. Good for children / adolescents > 8-10kg.
- Factors to control:
  - Ventilation: RR, Vt, Pi
  - Oxygenation: PEEP, FIO<sub>2</sub>, I:E ratio (1:2)

### SIMV:

- Ventilator detects patient breaths and waits until patient exhales to deliver mechanical breath.
- Ventilator delivers time-triggered assisted breath.
- If patient breathes between mandatory breaths, vent allows patient to breathe normal breath; may add some pressure-support for patient (non-vent) breaths. **Good weaning mode.** (Fig. 2)

### Bilevel / APRV: (Fig. 3)

- Good for severe ARDS. High level of continuous positive airway pressure ( $P_{high}$ ) that is maintained for a long period of time ( $T_{high}$ ) coupled with a short period ( $T_{low}$ ) during which the pressure is released ( $P_{low}$ ).
- The prolonged period of high pressure facilitates oxygenation and pulmonary recruitment.
- Need to have inverse I: E ratio.
- Oxygenation is often improved with APRV from increased mean alveolar pressure and volume from longer time above functional residual capacity or by creation of auto-PEEP.
- Patient **MUST** be able to breathe spontaneously (i.e. not be paralyzed).

## CPAP:

- Continuous pressure throughout the ventilatory cycle. Patient-triggered breaths with ventilator giving a constant pressure during inspiration and expiration. No pre-set rate, all patient's own breaths. No pre-set volumes. Good weaning mode.

## High-flow Oscillatory Ventilator:

- Lung-protective ventilatory mode. Often used when conventional modes failed. Reduces risk of volume-trauma. Alveoli stay inflated at a constant mean airway pressure. Oscillation prevents the lung inflate–deflate cycle. Good for oxygenation.
- o Factors to control: Mean airway pressure (keep low), frequency (Hz), Pressure amplitude, FIO<sub>2</sub>, bias flow
  - Mean airway pressure: Pressure to optimize lung volume and to increase the alveolar surface area for gas exchange. Recruits alveoli. Should initially be set at 2-3cm H<sub>2</sub>O above the corresponding mean airway pressure on conventional vent. Helps oxygenation.
  - FIO<sub>2</sub>: Same as on conventional ventilator; helps oxygenation
  - Amplitude: Variation around MAP. Determines tidal volume. Controls ventilation – increase the amplitude if under-ventilated. Normal amplitude 20-30 in neonates
  - Chest Wiggle: Seen from clavicles to lower ribs. Increased by increasing amplitude. Controls ventilation. When decreased / absent, may have mucous plugging, ETT disconnection/displacement, pneumothorax
  - Frequency: Controls ventilation. Decrease frequency if under-ventilated if amplitude is maximized

## High frequency Jet Ventilator:

- Through a pneumatic valve, releases short jets of gas in the inspiratory circuit; expiration is passive. Good for gentle ventilation of neonates (ex: CDH). Has reduced peak airway pressures.
- o Basic settings:
  - Rate: 420
  - Valve time : 0.20 seconds
  - PIP: 30 (or 20% increase from PIP on the conventional vent)
  - PEEP: MAP-4 on the conventional vent
  - Should do CXR 1h after placing patient on jet to look for lung expansion
- o Factors to control:
  - Ventilation: Change PIP or (rare) rate
  - Oxygenation: Changing FIO<sub>2</sub> or PEEP

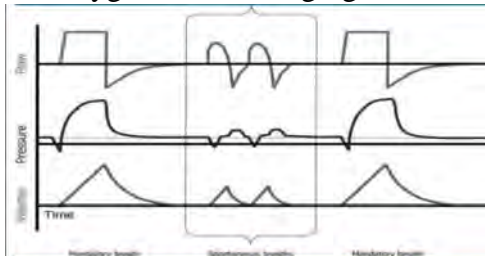


Figure 1

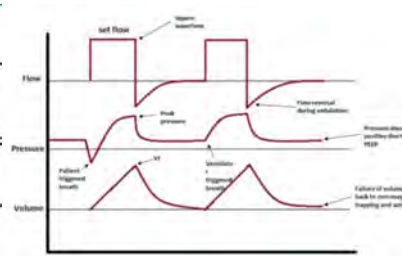


Figure 2

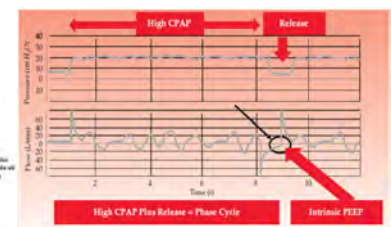


Figure 3

## REFERENCE FOR HASBRO NEONATAL AND PEDIATRIC ECMO

### Consults called for:

1. Meconium aspiration
2. Persistent pulmonary hypertension (PPHN)
3. Congenital diaphragmatic hernia (CDH)
4. Sepsis
5. Cardiac failure
6. Respiratory failure
7. If any doubt about indication, speak with on call ECMO attending or ECMO director

### Exclusion criteria

1. Less than 34 weeks gestation
2. Weight less than 2000 grams (relative contraindication)
3. Grade II or greater Intraventricular Hemorrhage
4. Irreversible organ injury (unless eligible for transplant)
5. Severe associated Anomaly (Trisomy 13/18) or other lethal chromosomal abnormality
6. Prolonged cardiac arrest
7. Unwitnessed or out-of-hospital cardiac arrest
8. On ventilator for greater than 10-14 days –chronic lung disease secondary to disease & iatrogenic injury
9. Tonic Clonic Seizures in newborns
10. Evidence of brain death or massive neurologic injury (exception: bridge to organ donation)
11. Uncontrolled bleeding diatheses or contraindication to anticoagulation

### Oxygenation index

Not as important as the trend of the blood gases, ventilator settings, pressors, urine output, but often communicated by the NICU and PICU. You must be at the bedside watching how the patient responds to titrations in the ventilator and drips.

Oxygenation Index greater than 35-45 on two or more blood gases is concerning for need for ECMO. **OI = (MAPxFiO<sub>2</sub>x100)/PaO<sub>2</sub>**

### Cannulation strategy

Should be calculated and documented in a note in the EMR even if that patient does not imminently need ECMO so that it can be referenced if the clinical status changes.

### Types of ECMO

Options include Veno-Arterial (VA) or Veno-Venous (VV) ECMO

- **Veno-Venous (VV)** – Purely respiratory problem, requires adequate heart function on echocardiogram
  - Need for pressors does not necessarily obviate the use of VV ECMO
- **Veno-Arterial (VA)** – Poor cardiac function, heart failure, cardiac arrest, septic shock

- Hybrid cannulations, i.e. VAV or VVA

## Cannula Size

- Calculation for full flow
  - Neonates less than 30 days – 150 cc/kg/min
  - Infants/children greater than 30 days – BSA x 2.4
  - Sepsis typically requires higher than usual ECMO flows, utilize the largest cannulas possible
  - For femoral cannulations would not recommend using an arterial cannula greater than 17 French due to risk of limb ischemia
- Look at chart (to follow) to see which size cannula can handle the calculated flow, have available cannulas that are one size smaller just in case

## Target vessels for cannulation

- Children <5 years old– Cannulate neck vessels (RIJ, RCCA preferable), femoral vessels are too small
- Children > 5 years & ~15kg– Cannulate femoral vessels (femoral artery/vein): this avoids ligating carotid artery if going on VA ECMO and increasing stroke risk
  - If enough time prior to cannulation, place arterial/venous central lines so that you can exchange over a wire to cannulas
  - Will need Distal Perfusion Catheter (DPC) – usually use 5F, coordinate with vascular surgery for placement early (even prior to cannulation)  
May consider using arterial cannulas in venous position, i.e. an arterial cannula in RIJ as a pull cannula especially in hybrid cannulations.

## Example of cannulation strategy to be documented in a note:

### CANNULATION STRATEGY:

#### VA ECMO

17 French Biomedicus arterial cannula to right femoral artery

21 French Biomedicus venous cannula to right femoral vein

5 French Distal Perfusion catheter to right SFA

Total flow: 4.3 L/min

Ht: 177.8 cm

Wt: 63.5 kg

BSA: 1.79 m<sup>2</sup>

Cardiohelp circuit with 3/8" tubing

### Steps to take and phone calls to make if proceeding with cannulation:

#### ECMO specialist (Respiratory shift supervisor): 401-255-3520

1. OR team: 401-444-5657
2. Blood bank: For infants, you will need 3 units PRBC to prime circuit. Call to blood bank can be made by PICU clerk. Have someone run to get the blood while RT ECMO specialists build the pump

3. Pharmacy: Place orders for meds for ECMO circuit and cannulation via the ECMO order set in Epic. Meds for pump can be overridden in the Omnicell. Order heparin bolus and make sure the PICU/ bedside nurse can administer.
4. PICU attending: should be “running the medical code” during cannulation. Titrating ventilator, medications/pressors/code meds, overseeing compressions if necessary if the cannulation is performed in the PICU.
5. Vascular surgery: if needing distal perfusion catheter (for ALL femoral cannulations) call and speak directly to the attending.

# ECMO Cannula Chart

VENOUS									
Central									
Biomedicus			Lighthouse				Right Angle Metal		
8fr	550cc/min	1/4"	.025 Wire	14fr	950 cc/min	12fr	900 cc/min		
10	1000	1/4"	.025 Wire	16	1300	14	1700		
12	1500	1/4"	.025 Wire	18	1850	16	2100 1/4" / 2000 3/8"		
14	2000	1/4"	.025 Wire	20	2400	18	2800 1/4" / 2800 3/8"		
15	1500	3/8"	.038 Wire	22	3000	20	3400		
15A	2100	3/8"	.038 Wire	24	3600	22	4400		
17A	2700	3/8"	.038 Wire	26	4400	24	5000+		
17	2100	3/8"	.038 Wire	28	5200	28	6000+		
19	3000	3/8"	.038 Wire	30	6000+	31	7000+		
21	4300	3/8"	.038 Wire	32	6000+				
23	5000	3/8"	.038 Wire	34	7000+				
**use BM pediatric perc kit for 8 -14fr**									
DLP		Avalon VV							
16A	2300	1/4"		13	600cc/min	1/4"	.038 Wire		
				16	900	1/4"	.038 Wire		
Origen VV				19	1500	1/4"	.038 Wire		
13	450	1/4"		20	1400	3/8"	.038 Wire		
16	850	1/4"		23	2200	3/8"	.038 Wire		
19	1050	1/4"		27	3400	3/8"	.038 Wire		
**use Origen dilator kit**									
				31	4700	3/8"	.038 Wire		
**use Avalon dilator kit**									
ARTERIAL									
Central					Peripheral				
Biomed.	Size	80% CPB BQ for long term							
640cc/min	<b>8fr</b>	1/4"	BM Peds Perc kit w/.025 wire						
1280	<b>10</b>	1/4"	BM Peds Perc kit w/.025 wire						
1800	<b>12</b>	1/4"	BM Peds Perc kit w/.025 wire						
2400	<b>14</b>	1/4"	BM Peds Perc kit w/.025 wire						
DLP	Flow								
16	3000	1/4"							
EOPA	Flow	**EOPA's can be used peripheral also**							
18	3900	3/8"	.038 Wire						
20	4700	3/8"	.038 Wire						
22	5700	3/8"	.038 Wire						
24	>5700	3/8"	.038 Wire						
Biomed.	Size	80% CPB BQ for long term							
640cc/min	<b>8fr</b>	1/4"	BM Peds Perc kit w/.025 wire						
1280	<b>10</b>	1/4"	BM Peds Perc kit w/.025 wire						
1800	<b>12</b>	1/4"	BM Peds Perc kit w/.025 wire						
2400	<b>14</b>	1/4"	BM Peds Perc kit w/.025 wire						
3500*	<b>15</b>	3/8"	.038 wire						
4200	<b>17</b>	3/8"	.038 wire						
5000	<b>19</b>	3/8"	.038 wire						
>5000	<b>21</b>	3/8"	.038 wire						

## PEDIATRIC NUTRITION

### NUTRITIONAL REQUIREMENTS

The hospital and JCAHO **mandate** that all patients be screened for nutritional status after three days (ICU patients sooner).

- You will see these screening notes from Nutrition Services on the chart and the dietitian may speak to you about their concerns.
- **These must be relayed to the fellow or service attending for discussion.**

Consultations for nutritional support from the GI service are only obtained upon attending request. However, the **intestinal failure** patients are now managed by a multidisciplinary team consisting of Surgical staff, GI, Nutrition, Social Work, OT, PT, and Speech. **All intestinal failure patients (short gut) who are admitted will require consultation for all these services.**

#### Caloric requirements:

0-1 year 90-120 kcal/kg/day

1-7 years 75-90 kcal/kg/day

7-12 years 60-75 kcal/kg/day

12-18 years 30-60 kcal/kg/day

- Physical status (metabolic condition, bedridden, active) will affect these baseline values.
- For infants, the basic principle is that sufficient calories need to be supplied to assure continued growth and development.
- For chronically hospitalized infants it is important to **maintain an accurate growth record in EPIC**. If weights are ordered they will be populated in an age-appropriate growth chart. Please order daily weights for admitted patients, especially infants and those with ongoing illnesses.
- o The expected weight gain is 1% of the infant's weight per day. i.e. a 2 week old, 1kg, 34 week gestation infant should gain about 10 g/day.
- o In very small infants it can be difficult to achieve a certain caloric intake without exceeding fluid intake limitations. For that reason, both ml/kg/d and kcal/kg/d should be calculated daily.

## PARENTERAL NUTRITION

### Indications for Initiation of Parenteral Nutrition

Prematurity: Required within 24-48 hour of birth secondary to low nutrient reserves, increased energy expenditure, immaturity of the GI tract and increased risk for acute or chronic illness

### Gastrointestinal conditions potentially requiring Parenteral Nutrition:

#### Congenital

Esophageal/intestinal atresia

Tracheoesophageal fistula

Malrotation with volvulus

Hirschsprung's disease

Anorectal malformation

Abdominal wall defects

#### Sort term and long term GI disorders

Severe inflammatory bowel disease

Pseudo-obstruction/obstruction

Motility disorders

Acute pancreatitis

High output fistulas

Necrotizing enterocolitis

Short bowel syndrome

#### Critical illness

Hemodynamic instability

Enteral feeding intolerance

Chyle leaks if unresponsive to enteral feedings

#### Oncology

Graft vs host disease

Radiation enteritis

Cachexia

#### Miscellaneous

Organ failure (If enteral nutrition is contraindicated and in the setting of catabolism or in preparation for transplant)

## TOTAL PARENTERAL NUTRITION (TPN)

When writing TPN orders, there are some additional principles to adhere to beyond the simple provision of calories. We can divide this conceptually, into the provision of carbohydrate, protein and fat intravenously.

1. Carbohydrate: The **amount** of dextrose supplied per day, the **caloric density** and particularly the **rate** at which it is administered are critical.

- The caloric density of carbohydrates in solution is 3.4 kcal/g glucose.
- From this one can derive the caloric density of any % dextrose solution.
- Example: D10W solution has 10g glucose/100 ml and provides  $(10\text{gm} \times 3.4\text{kcal/g})/100\text{ml} = 0.34\text{kcal/ml}$ ; D20 W solution has twice the caloric density, 0.68kcal/ml. The other solutions' density can be similarly calculated:
- D5W 0.17 kcal/ml; D10W 0.34 kcal/ml; D12.5W 0.42 kcal/ml; D15W 0.51 kcal/ml; D17.5W 0.59 kcal/ml; D20W 0.68 kcal/ml; D25W 0.76 kcal/ml D30W 0.85 kcal/ml
- Peripheral TPN (rarely used in surgical patients) **should not exceed** D12.5W or 900 mOsm/L and central TPN **should not exceed** D20W, unless there are severe restraints on fluid intake.
- D25W - D30W have osmolarities of between 1200 and 1700 mOsm/L. These solutions **are useful for patients on ECMO** support where not only are the fluids restricted, but the large cannula in the atrium also allows for prompt dilution of the infusion.

### TPN order sets

- TPN order sets are provided in EPIC.
- They are weight based and offer a standard admixture of macro and micro-nutrients based on age groups (infant, child, adolescent).
- They are also displayed as Cyclic TPN for the infant, child, and adolescent.
- Lipid orders are linked to each and the prescriber may choose TPN with standard lipid emulsions or SMOF lipids. These will be described later in this text.

## Indications for initiation, advancement of macronutrients in parenteral nutrition

### Dextrose Requirements:

When initiating TPN, the **rate** of glucose provision should be increased incrementally.

- In neonates and premature neonates this requires calculation of total grams of glucose provided per day and at an initial rate probably not exceeding **6.5mg/kg/hr**.
- For older infants and children this can be accomplished most easily by starting with **D10TPN** and progressing to D20TPN over about 3-4 days, while providing it at rates sufficient to support maintenance fluid requirements.
- Alternatively, in an older child one can start with D20TPN, but infuse it at a rate 25% of the ultimate goal rate and increase by 25%/day. This requires other fluids to be infused concurrently to meet fluid requirements. (This may not be feasible when IV access is limited and you have a line that you want to devote to TPN only).
- To monitor tolerance of the dextrose infusion, urine should be dipped for glucose and bedside blood glucose monitoring should be obtained every 6 – 8 hours until 24 hours at the goal rate have been attained. Then monitoring decreases to once a day.

### Fat Requirements:

Fat is used as an energy source and to provide essential fatty acids (linoleic and linolenic acid). With a caloric density of **9 kcal/g**, it is a valuable source of calories in parenteral nutrition.

- 20% intralipid (20g fat/100ml fluid) solution has a caloric density of 2kcal/ml.
- Normally **30-40%** of total calories are provided by fat in the form of 20% intralipid solution. It is generally undesirable to exceed 30% of calories.
- Exogenous fat is handled and metabolized in the same manner as that presented as chylomicrons. **Lipid emulsions should therefore be infused cautiously.** The enzyme lipoprotein lipase can be saturated by an excessive infusion, which results in lipidemia.
- Guidelines for infusion of 20% intralipid(g/kg/day) for premature to term infant:

Initial 0.5-1 grams per kg/day. Increase daily by: .025-0.5 gm/kg/d, Max dose: 3 gms/kg/d. Monitor the effect of infusion by measuring the serum triglyceride level. The goal is <200mg %.

\*\*Caution should be used in infusing fat emulsion in all patients with pulmonary insufficiency, liver failure, jaundice and coagulation disorders. In older children, an allergy to egg whites is a contraindication to use of intralipid.

Standard lipid emulsions are derived from Soy. There are various forms of lipid now available. SMOF is a combination of soy, MCT, Olive oil, and Fish oil. Omegaven is derived from Fish oil. Both SMOF and Omegaven have been reserved for long term TPN patients due to their anti-inflammatory properties which are believed to mitigate/prevent the onset TPN cholestasis

### Protein Requirements:

The amino acid composition of neonatal, childhood and adult TPN differ as there are some amino acids that appear to be conditionally essential during the early phases of life. Further changes occur when there is impairment of either renal or hepatic function.

The calories that could potentially be derived from these solutions are not calculated into the calorie provision. Protein requirements for growth and repair must be met and are generally age dependent. (See table)

#### Protein requirement (g/kg/day)

- Premature-term neonate 0 to 1 month: 3.0-3.5
- infant 1-12 months: 2.5-3.0
- children 1-12 years: 1.5-2.5
- adolescents 12-18 years: 1.0-1.5

The "**total calorie**" to **nitrogen ratio** of the TPN formulation has great impact on the optimal utilization of the carbohydrate calories and potentially on the incidence of TPN associated liver disease. Ideally it ranges from 150-180:1. In order to calculate this ratio you must calculate a total number of kilocalories provided by both the dextrose and fat emulsion over one day and divide it by the grams of nitrogen infused with that same volume of TPN. EPIC calculates the Nitrogen to non- nitrogen calorie ratio and it appears in the left margin of the order set.

An **example** for calculating the Nitrogen to non-nitrogen calorie ratio is as follows:

An infant is receiving 500 ml/day of TPN, which consists of D20 solution and 50ml of 20% intralipid. He is also receiving 3g/kg/d of protein. His weight is 5 kg.

- $500\text{ml} \times 0.68\text{kcal/ml} = 340 \text{ kcal/day from TPN}$

- $50\text{ml} \times 2\text{kcal/ml} = 100 \text{ kcal/day}$  from intralipid
- $3\text{g/kg/day} \times 5\text{kg} = 15 \text{ g protein}$
- conversion to nitrogen grams:  $6.25\text{g protein} = \text{one gram Nitrogen}$
- $15 / 6.25 = 2.4 \text{ g Nitrogen}$

Total calorie: Nitrogen ratio =  $(340+100)/2.4 = 183 :1$

This same example serves to illustrate how ml/kg/day and kcal/kg/day are calculated.

The total volume infused is  $500 + 50 = 550\text{ml/d}$ . For a 5kg infant this is  $110\text{ml/kg/day}$ .

The caloric provision is  $340\text{kcal/day}$  from TPN and  $100\text{kcal/day}$  from fat. Divided by 5kg yields  $88\text{kcal/kg/day}$ .

These components of TPN exist in infant and pediatric formulations and are ordered on a per kg basis. There are specific instances when these substances need to be reduced or even withheld. This will be indicated by the fellow or attending monitoring the TPN orders in collaboration with Nutrition and Pharmacy services.

An example for calculating goal TPN/Intralipid volumes for an infant or child:

You are asked to start TPN for a 20kg child.

- -The estimated caloric need is **80 kcal/kg/day**.
- -The total fluid rate will be approximately **20%** higher than this number, i.e.  $80+16=96\text{ml/kg/d}$   
That would be  **$20\text{kg} \times 96 = 1920\text{ml/d} = 80\text{ml/hr}$**
- -10% of this rate should be the intralipid rate and the other 90% should be TPN infusion rate.

IL @  $8\text{ml/hr} = 192\text{ml/d} = 384\text{kcal/day}$

TPN @  $72\text{ml/hr} = 1728\text{ml/d}$ ; if D20 then  $1726\text{ml} \times .68\text{kcal/ml} = 1175\text{kcal/day}$ ;

Total kcal/day =  $1175+384 = 1559$  divide by  $20\text{kg} = 78\text{kcal/kg/d}$ .

If the child is getting close to  $3\text{g/kg/d}$  of protein in the TPN, then the cal:N ratio will be 162:1. Remember that these are **the goal** concentrations/rates for infusion and **not the initial ones!** For instance: initiate D10TPN at  $72\text{ml/hr}$  and 20 % IL at half the goal rate  $4\text{ml/hr}$ . The next day advance to D15TPN at  $72\text{ml/hr}$  and increase IL to  $6\text{ml/hr}$ , then advance to D20TPN at  $72\text{ml/hr}$  and IL at  $8\text{ml/hr}$ , if glucose and lipid tolerance are verified. (Dextrostix and triglyceride levels). Careful monitoring of fluid status and weight gain will allow for subsequent adjustments.

### **Micronutrients**

As earlier described the Pediatric TPN order sets cover the infant, child and adolescent and are weight based. The order sets for each age group come with standard electrolyte dosing for each age group based on daily requirements. These doses may be altered based on deficits and ongoing losses. Electrolytes within TPN may be adjusted to offset these deficits but caution should be taken to avoid repleting electrolytes using TPN unless the corrections are minor. It must be noted that orders placed for TPN on a given day will not be administered until 9 pm that night so any corrections/responses to repletion using TPN orders will be delayed. The following are guidelines for dosing parenteral nutrition electrolytes and multivitamin. Dosing for trace elements is standardized and is usually not individually dosed except during times of drug shortages. The pharmacy staff should guide you in making those decisions.

<b>Electrolyte</b>	<b>Preterm Neonates</b>	<b>Infants/Children</b>	<b>Adolescent &gt;50kg</b>
Sodium	2-5 mEq/kg	2-5 mEq/kg	1-2 mEq/kg
Potassium	2-4 mEq./kg	2-4 mEq/kg	1-2 mEq/kg
Calcium	2-4 mEq/kg	0.5-4 mEq/kg	10-20 mEq/day
Phosphorus	1-2 mmol/kg	0.5-2 mmol/kg	10-40 mEq/day
Magnesium	0.3-0.5mEq/kg	0.3-0.5 mEq/kg	10-30 mEq/day
Acetate	As needed for acid base balance		
Chloride	As needed for acid base balance		

### Parenteral Vitamins

< 1 kg	1.5 mL
≥1 to <3 kg	3 mL
≥3 kg	5 mL

### Cyclic TPN

Cycling TPN is a maneuver to further reduce the pathophysiologic consequences of TPN administration. It needs to be achieved gradually and the usual goal is to infuse the necessary volume of TPN and intralipid over a **16 – 18 hour period**. One hour at each end of the infusion time will have the solution running at half the normal rate to allow the pancreas to adjust insulin/glucagon secretion to prevent hypo- or hyperglycemia. EPIC will calculate the ramp up and ramp down one hour before and after TPN is cycled off. You must use the Cycled TPN order sets for cycling TPN. Bear in mind that infants and very young infants may not tolerate long periods of time off TPN unless a sufficient volume of enteral feeds have been initiated to offset nutritional requirements that are absent when cycled off. Older children may be cycled off for 8-12 hours once stable and in preparation for home nutrition support.

To calculate these rates divide the total volume of TPN/day by the number of hours for infusing minus one to obtain the rate of infusion during the main period of infusion. One half that rate is the step-up and step-down rate.

Example: for the previous 20-kg child:

- The 1920cc volume is to be infused over 18 hours.
- $1920 / (18-1) = 113\text{cc/hr}$
- Start the infusion at 56cc/hr x 1 hr, then increase to 113cc/hr x 16 hours, then decrease to 56cc/hr x 1 hr, then stop TPN for 6 hours before resuming the cycle.
- Intralipids are not tapered, so the rate of the intralipid will be  $192/18 = 10.6\text{cc/hr} \times 18\text{hrs}$

Running D10 solution or other fluid should be unnecessary if the patient has been gradually tapered to this regimen from a 24-hour infusion protocol. This is a lot of calculator time, but some of this is calculated within the EPIC platform. The purpose of providing the calculations to you is to give you a better understanding of the complexity of TPN administration.

### Frequency of Short-and Long-Term Metabolic Monitoring

CMP Magnesium and phosphorus- Initially monitored daily and modified to three times weekly, then weekly when stable. Once stable and transitioned to long term TPN, these labs may be

repeated every other week. However, during acute illness or hospitalization these parameters may be monitored more frequently even in a long-term patient who has been stable on TPN.

Prealbumin, Triglycerides, CBC, LFTs and GGTP- Initially weekly and monthly once stable

Iron studies, Zinc, Selenium, Manganese, Copper Chromium Carnitine- Initially every 3 months and every 6 months once stable

Fat soluble vitamins, B12- Initially every 3 months and every 6-12 months once stable.

**Note:** While in the hospital setting blood drawing from a central line used for TPN should be kept to a minimum and care should be taken to avoid breaking into a line while the TPN is infusing. Our general protocol is to avoid blood sampling until the new TPN is hung or the TPN from the previous day is taken down in the evening hours except when results are otherwise clinically advisable (Drug levels, blood cultures for temperature spikes etc.)

## ENTERAL NUTRITION

### A simple approach to infant formulas

Multiple formulas attempt to provide nutrients in a similar composition to human breast milk. The primary components of any nutritional product are the 1) protein source, 2) carbohydrate source and 3) fat source. The manner in which these components are supplied is the chief difference between the various infant formulas and breast milk. The osmolarity of the solution and electrolyte composition have additional and significant effects; these components must be considered in the enteral nutrition of the premature newborn, in particular. Most formulas are either premixed as a liquid concentrate or a powder that is reconstituted with water; reconstitution permits changes in the caloric density (and osmolarity). This form of titration can be used for dilution of feedings in order to reduce the osmolar load when starting feeds. The caloric density for breast milk and infant formulas unless specifically noted is  $20\text{kcal/ounce} = 20\text{kcal}/30\text{cc} = 0.68\text{kcal/cc}$ .

The list below is a simple description of the most commonly used formulas; there are many more highly specialized formulas on the market that are described elsewhere.

### Infant formulas

#### Milk based formulas:

**Similac, Enfamil, Good Start:** The protein source is from non-fat milk, whey or casein; can also be milk protein isolate for patient with intolerance. The fat source is primarily coconut and soy oils. The Protein:Fat:CHO ratios are about the same and the osm for 20kcal formula is 300 mOsm/kg of H<sub>2</sub>O. Very similar products from different manufacturers.

#### Lactose free formulas:

**Isomil, Prosobee and Similac Soy:** Sucrose or corn syrup solids are the CHO source. The protein source is soy protein in all three and the fat source is coconut, soy or safflower oil. Again, similar products from different manufacturers.

#### Protein hydrolysate formulas (for infants with absorption problems):

**Alimentum, Nutramigen, Pregestimil and Portagen:** These are all casein hydrolysates. Their CHO and fat sources vary. There is significant variability in the osmolarity in these formulas, with Pregestimil being closest to normal. Alimentum, because of its higher osmolarity, needs to be used with caution. Nutramigen differs from Pregestimil and Portagen in that the latter two are appropriate for situations in which the long chain fatty acids need to be limited. However, Portagen provides lactose, which may be a problem for some infants.

#### The Premie formulas:

**Enfamil Premature, Similac Special Care, Similac Neosure:** These formulas are lactose based with other carbohydrate sources, the protein source is whey/casein and the fat source is a combination of MCT/soy safflower, sunflower and coconut oils. They differ from their regular infant formula counterparts in that they have 1) other sources of CHO, 2) twice-higher amounts of sodium and calcium per 100 ml and 3) slightly more iron. Please note: mixing the regular infant formulas to a higher strength still does not provide the necessary electrolytes and significantly raises the osmolarity. This has particular implications in the setting of malabsorption as in the case of intestinal failure.

(24kcal Similac has 380mOsm/kgH<sub>2</sub>O vs 24kcal Similac Special Care which has 300mOsm)

### Free Amino Acid Formula

**Elecare Infant, Neocate Infant:** These formulas are elemental. The protein source is free amino acid or free L-amino acid. The CHO source is corn syrup solids and the fat source is primarily a combination of high oleic safflower oil, MCT, refined vegetable oil, and soy oil. These formulas are primarily reserved for conditions associated with malabsorption or milk protein allergy.

### **Toddler Formulas**

After the first year of life as Cow's Milk had previously been avoided until after the first year. However recent recommendations from the American Academy of Pediatrics suggest cow's milk may be introduced before one year of life.

The composition of formulas for children over one year who are dependent on tube feedings or non-solid nutrition also changes. Typical formulas include **Pediasure, Isocal, Osmolite, and Ensure**. Again, CHO and protein as well as fat sources differ. They have a higher caloric density, usually 30kcal/oz (one kcal/cc) and come in low or high residue varieties (containing fiber). These products vary in complexity. There are polymeric enteral products that include **Boost kid Essentials, Boost Kid Essentials 1.5 (1.5 kcal/mL), and Compleat Pediatric** (composed of "real food") in a liquidized form. Peptide based pediatric formulas include **Peptamin Jr. Peptamin 1.5 and Vital Jr**. Some of the more palatable formulas are used as supplements that are taken by mouth when children are unable to tolerate enough nutrition from an age-appropriate diet to meet demands for growth.

### **Enteral Nutrition in Pediatrics**

#### Role of Enteral Nutrition

In patients with a functional and safely accessible gastrointestinal tract, enteral nutrition is preferred over parenteral nutrition. It supports normal endocrine, paracrine, and neural function. It improves mesenteric blood flow, decreases permeability within the gastrointestinal tract and prevents structural and functional alterations of the gut barrier. Enteral nutrition also plays a role in the promotion of pancreatic and biliary secretions. In addition, it lowers the risk of metabolic complications such as glucose and electrolyte abnormalities. Lastly, it reduces infection risk, lowers cost of nutrition support, and promotes gut-related immune system function.

Indications for Enteral Nutrition. The following indications include but are not limited to:

***Inadequate oral intake for demand***

- Cystic Fibrosis
- BPD
- CHD
- Renal disease
- Infection
- Surgery
- Burn, Trauma
- Failure to thrive

***Developmental***

- Preterm birth
- Prolonged intubation in a neonate or infant
- Neuromuscular disorders
- Neurological impairment

***Behavioral***

- Feeding aversions
- Unpalatable diet
- Eating disorders

***Anatomical***

- Cancer, burns, mucositis
- Congenital malformation
- Tracheosophageal fistula

***Airway Protection***

***Anatomical***

- Tracheosophageal fistula
- Paralyzed vocal cord
- Severe reflux requiring postpyloric feeding
- ***Developmental***
- Neurodevelopmental delay
- Cerebral palsy

***Inadequate Intestinal Function***

***Malabsorption***

- Intestinal failure (short bowel syndrome)
- Hi output ostomy
- Crohn's disease
- Pancreatic insufficiency

**Tube Feeding Methods**

Method of delivery depends on the child's age, condition, diagnosis and placement of the feeding tube

Intermittent bolus feeding: More closely approximates normal physiologic GI function (gastrocolic reflex)

Continuous infusion: For severe malabsorption, dysmotility disorders, or in conditions where the patient does not tolerate peaks and troughs of osmolarity associated with bolus feeding.

Combination of intermittent and bolus: May be used when transitioning from continuous to bolus feeds

Post pyloric/Enteric: For infants with severe reflux, delayed gastric emptying, or both

*Methods for Advancement of Continuous and Bolus/Intermittent Feeds*

The following are guidelines only and advancement may be highly individualized, dependent on anticipated tolerance and based on the child's responses and underlying condition. It is recommended that you make one change at a time when adjusting feeds both in volume and concentration and the patient should be monitored to assess for feeding intolerance (distended abdomen, emesis, diarrhea, cramping etc.). Bolus feeds are not recommended in post pyloric feeding tubes.

***Bolus***

In children **up to one year** unless otherwise indicated, bolus feeds may be started at 10-15 mL/Kg every three hours. Feeds may be advanced by 10-30 mL per feed as tolerated to goal.

In Children **one year to 6 years**, start at 5-10 mL/kg every three hours. Feeds may be increased by 1mL/kg every feed or every other feed as tolerated to a maximum of 15-20 mL/kg every 4-6 hours

Children **7 years and up** may be started at 25 mL/hr and increased by 25 mL every feed or every other feed as tolerated with a maximum of 330 to 480 ml every 4-6 hours.

***Continuous***: May be indicated in delayed gastric emptying, risk for aspiration, malabsorption.

Children **up to one year of age** may be started at 1-2 mL/kg/hour and advanced by 1-2 mL/kg every 3-8 hours based on tolerance and not to exceed 6 mL/kg/hour.

Children from **1-6 years of age** may be started at 1 mL/kg/hour and increased by 1mL/kg every 3-8 hours not to exceed 5 mL/kg/hour.

Children **7 years and up** may be started at 25 mL/hour and advance by 25 mL every 3-8 hours not to exceed 100-150 mL/hour.

*Enteral and Parenteral Access will be discussed in another section.*

## Ketogenic diet (KD) guidelines when NPO and for procedures and surgery

### Goal while NPO:

Maintenance of Ketosis: Beta hydroxybutyrate 3-6 mmol/ L

Maintenance of blood glucose levels: above 50 mg/dL (typically 50-80mg/dL)

Maintenance of CO<sub>2</sub>  $\geq$  18 mmol/L

**Fasting:** The ketogenic child can be fasted in the same fashion as other children. Clear liquids on the KD means, “sugar-free, caffeine free”, therefore diet caffeine-free sodas, sugar-free gelatin, ice chips, half strength Pedialyte (half Pedialyte + half water) and water are allowed.

**Medications:** Must be in lowest carbohydrate form such as crushed swallow tablets or I.V. form (no chew tabs, syrups, or elixirs). Check with pharmacy before ordering new medications.

**Intravenous solutions:** Use 0.9 NaCl (normal saline). Lactated ringers are also carbohydrate free. Dextrose solutions should be avoided unless fasting for greater than 12 hours when it may be necessary to maintain stable glucose levels

**Glucose levels:** Check prior to surgery and every 6 hours until stable. If blood sugars drop below 50 mg/dL, give 30 ml juice and if NPO, IV bolus of 50 ml D5W (no continuous dextrose) over 30 minutes. Recheck again in 45 minutes and if still low, repeat. Glucose levels usually increase due to stress response (due to increased cortisol levels). \*\*Usual glucose levels for ketogenic patients are 50-80 mg/dL.

**CO<sub>2</sub>:** Check every 12 hours with basic metabolic panel. Extended fasting may lead to excessive ketosis and acidosis. Please make sure patient is receiving home regimen for sodium bicarbonate supplements. If not able to take by mouth, intravenous bicarbonate should be administered to correct acidosis (If CO<sub>2</sub> < 18 mmol/L).

### General sodium bicarbonate dosing guidelines:

2-3meq/kg/day given continuously with IV fluids or in divided doses of 2-3 times daily.

If 2-3meq/kg/day, or home dosing regimen does not provide adequate repletion and CO<sub>2</sub> continues to be <18mmol/L and or patient is symptomatic increase supplementation starting at 50% of calculated deficit and provide as continuous parenteral infusion:

**Fluid volumes:** Children on the KD follow a maintenance “fluid schedule” to ensure adequate hydration throughout the day. There have been reported cases of children on the KD who have had breakthrough seizures after receiving large volumes at one time, therefore avoid acute over-hydration.

Maintenance fluid calculations (Holiday-Segar method):

1-10 kg	100 ml/kg
11-20 kg	1000 ml + 50 ml/kg for each kg >10 kg
>20 kg	1500 ml + 20 ml/kg for each kg >20 kg

### Advancement of diet (based on clinical discretion):

Post-surgery

1. First 2 hours: ½ Strength Pedialyte- 1 part water to 1 part Pedialyte at ½ maintenance
2. Second 2 hours: ½ Strength Pedialyte- 1 part water to 1 part Pedialyte at maintenance

### Initiation of keto formula and solids:

Start at half volume keto formula and ½ serving of keto meals  
Advance per clinical discretion to full volume of ketogenic formula and full serving ketogenic meals

Jeannie Dorr, MS, RD, LDN, 401-350-2808 or 401-444-8491

Jessica Mulligan, MS, RD, LDN 401-350-5254 or 401-444-8492

## VASCULAR ACCESS

- Vascular access is an important part of the management of Pediatric Surgery patients for resuscitation, phlebotomy, medication, and other indications
- Most commonly vascular access is established via a peripheral IV.
- Other forms of access may be required if:
  - It is expected that the patient would have a prolonged hospitalization (>7 days)
  - The patient will require ongoing medical management in the outpatient setting (i.e. antibiotics or parenteral nutrition)
  - The patient is reported to be a “difficult access” or requires frequent phlebotomy

### Peripherally Inserted Central Catheter (PICC Line) (Brands: Bard)

- Placed in a peripheral vein with the catheter terminating most commonly in a central vessel (SVC/IVC).
- Placement is performed with sterile technique at the bedside.
- Done by direct visualization/palpation, transillumination or ultrasound guidance
- Classified as a midline catheter or a central line.
- **Important:** the location of the termination of the line will direct what therapies can be infused.
- Can be single, double or even triple lumen.
- Hasbro Children’s Hospital’s PICC line team is also its Pediatric Sedation team.
- Consults can be placed in Epic and the team can be contacted at 4-6091.
- Will need another call to Pediatric Sedation by an APP, Fellow, or Attending to confirm request
- When asking for a PICC line consult, establish with the PICC team the indication for the consult as well as how many lumens are preferred.

### Non-Tunneled Central Line (Brands: Cook, Arrow).

- Can be placed either at bedside or in the OR.
- Sterilely placed by either ultrasound guidance or anatomical landmarks, sutured in place.
- Stable vascular access for duration <2 weeks for patients who are not PICC Line candidates for various reasons or are too critically ill to tolerate a trip to the OR
- As these lines are sutured in place and the catheter can be as short as 5cm in length, **preservation of these lines is paramount.**
- Daily assessments to ensure intact sterile dressing and sutures are needed

### Tunneled Central Lines.

- Placed in the OR
- Used for patients who require long term central access
- Vascular access obtained with ultrasound guidance or anatomical landmarks.
- After accessing the vessel, the catheter is tunneled under the skin away from the needle site; commonly exits in the pectoral region.
- Fluoroscopy is used to correctly position tip, usually in SVC or at SVC/RA junction
- Sutured in place at time of placement to help preserve the line

- Implanted cuff on the catheter is subcutaneous and over time will adhere to surrounding tissue to maintain placement and prevent dislodging
- Removal requires disrupting/mobilizing the cuff and holding pressure at the **ENTRY** vessel, not the entry site of the catheter
- Can be done in the OR or at the bedside/sedation suite for some patients
- Common names: “Hickman®,” “Broviac®,” “Tunneled Central lines”

### Port-a-cath

- Central venous catheter completely implanted under the skin with a pierceable reservoir
- Silastic top of reservoir allows for needle access
- Silastic tube travels under skin from reservoir to major vein
- Used for chemotherapy and other chronic treatments
- Can also be used for blood draws
- Placed in OR with image guidance or anatomical landmarks
- Vessel cannulated, catheter tunneled from the reservoir site where it has been connected
- Removal requires OR
- For children >10kg due to size and equipment limitations, although sometimes placed for smaller patients depending on the circumstances. Check with attending

### Consult a for central line

When another service requests a consult for central line placement, the following must be established:

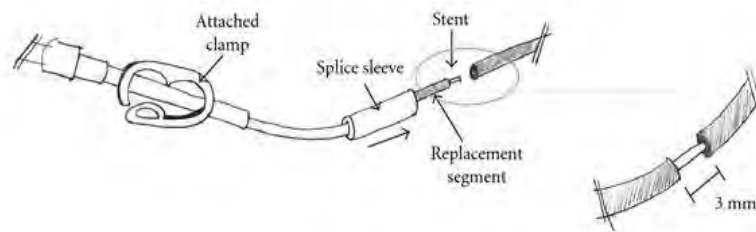
1. Indication for consults
2. Duration that line will be needed
3. Existing access which the patient has in place
4. How many lumens the requested line should have
5. If the consult is from the Pediatric Hematology/Oncology team:
6. Establish indication/diagnosis for the request for vascular access
7. Determine if the request is for a port or a Broviac, and the number of lumens needed
8. The patient and/or family must be aware of the diagnosis from the primary team and be anticipating a surgical consult. This is the expectation before any member of the pediatric surgery team will see the patient to perform the consult.
9. Any patient with a blood-based malignancy should have a CXR to check for a mediastinal mass. Also inquire about difficulty breathing or orthopnea
10. Check patient labs, especially ANC and platelet count, and be ready to discuss results with attending when presenting consult.
11. There **MUST** be clear documentation by the primary team, signed by the Heme/Onc attending, attesting to the type of line (port vs. Broviac) prior to going to the OR for placement.

Note: often the consult for a new oncology diagnosis will be Attending to Attending.

## Trouble-Shooting Central Lines

### Repair of a Broken Tunneled Central Line

Surgery is often consulted for a “broken Broviac” or “broken tunneled central line (CVL).” In certain settings the catheter can be repaired at the bedside. Timely interventions when the damage is discovered (clamp catheter (hemostat over sponge to prevent breakage), protect from contamination) will be done by the consulting team but a Pediatric Surgery team member should immediately see the patient and assess that these steps have been taken. Repair kits can be found in the PICU, on the 4<sup>th</sup> floor and 5<sup>th</sup> floor as well as the ED and Central Supply. Repair kits come in sizes 4F, 7F, 9F, and also as single and double lumen. The Pediatric Surgery team member must verify the correct size repair kit for the damaged line.



### Repair process

1. Establish the catheter is clamped with no active bleeding or fluid leakage from damaged area.
2. Establish the correct size repair kit to be used.
3. Note what type of damage is present: dissection, tear, hole, ballooning.
4. At least 5cm of the catheter distal from the site of insertion into the skin must remain; if it is a double lumen with one broken lumen, at least 2.5 cm should remain distal to the bifurcation.
5. Under sterile technique, the sterile occlusive dressing should be taken down and site cleaned thoroughly.
6. The new catheter in the repair kit should be flushed with normal saline. Glue within the kit should be prepared in provided syringe.
7. Proximal to area of damage, the old catheter should be cut and removed from table.
8. The new catheter should be inserted and applied with some force to establish silastic portions line up with no visible gaps (the metal of the stent should not be seen). The sliding sleeve should be placed over the area of repair. Glue should be generously administered through both ends of the sleeve and distributed under the sleeve so that the repair is completely encompassed with glue.
9. Hold the repair together for a few minutes to allow the glue to cure. Alternatively, a splint such as a tongue depressor or IV board can be secured along the sleeve joint to better protect the repair; this can stay in place for up to 48 hours. A new sterile dressing should be applied at the earliest opportunity.
10. Draw back to ensure blood return, then gently hand inject antibiotics through the repaired line (preferred: Ancef 25 mg/kg over 3-5 minutes).

11. After completing the antibiotics, again draw back to ensure blood return the gently flush with saline. Next, gently flush the line with 10units/1ml of heparin and leave the line heparin locked for 4 hours before using.
12. If the patient has a double lumen, blood cultures should be obtained, and prophylactic antibiotics should be administered through the non-affected lumen. If single lumen, blood culture and antibiotic should be done at the conclusion of 4 hours. At conclusion of repair, the provider should leave a note describing the procedure and how well it was tolerated. If the patient is afebrile, they can be discharged home at this time.

## Repair Kits: Inventory and Location

All repair kits can be found in Central Supply. When asking for a kit, it best to use the locator/inventory number to identify the kit.

### Double Lumen Kits

5 Fr #107915

6 Fr #156260

7 Fr #378391

### Single Lumen Kits

4.2 Fr #1788

9.6 Fr #1789

Larger kits (which are rarely used) can be obtained from Adult Oncology.

A limited number of single lumen kits can be found in the HCH 4 and 5 treatment rooms (double lumen repair kits are not stocked on the floor)

## TPA/Alteplase/CathFlo

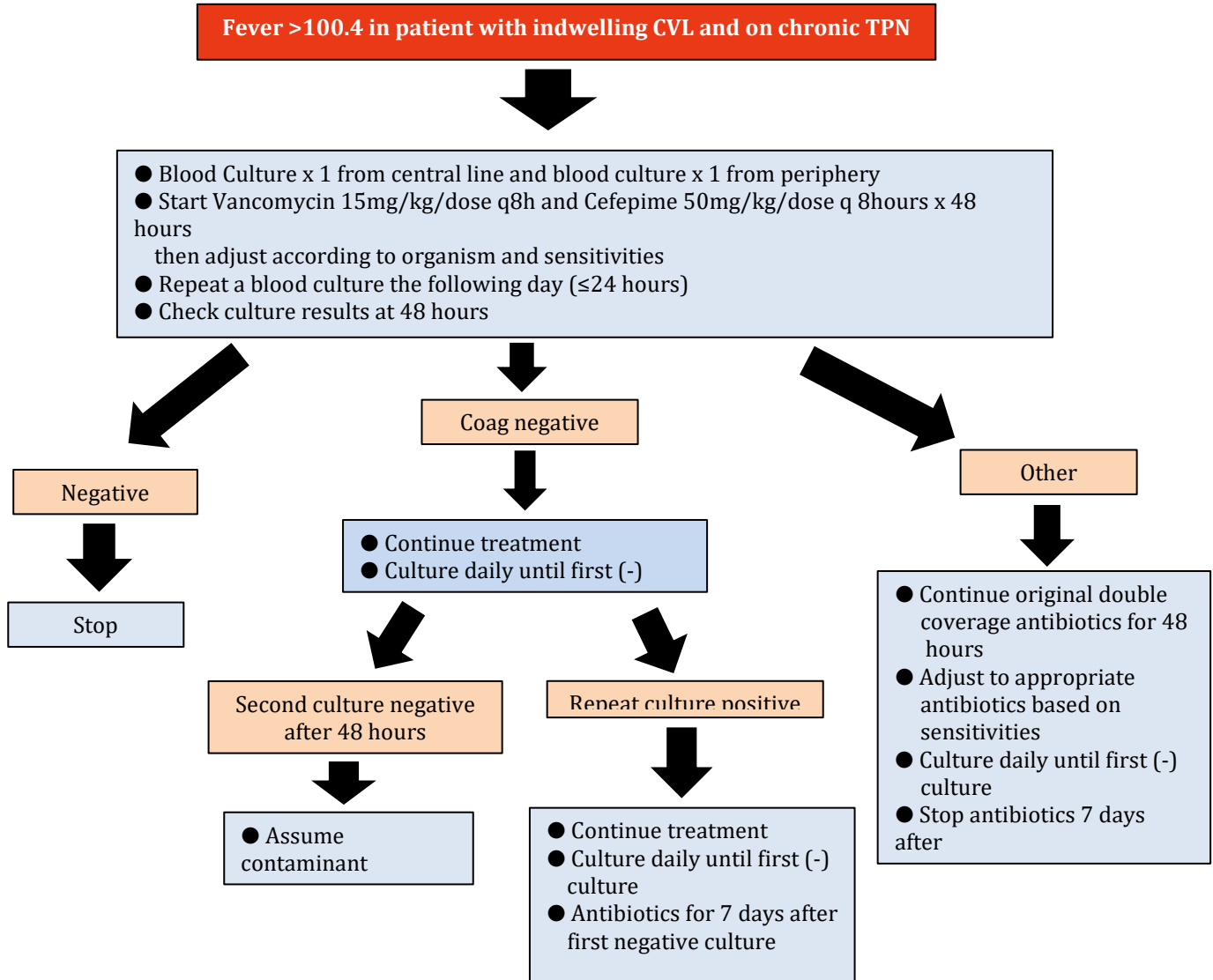
Central/Midline catheters are at risk of thrombotic events with occlusion of the catheter. This results in sluggishness or inability to infuse and/or lack of blood return. Alteplase/CathFlo can ordered and administered by nursing. Alteplase/Cathflo is a specific order in Epic and standard dosing for catheter clearance is 1mg (1mg/1ml). This will be instilled by the bedside RN. The dwelling time should be 90 mins, after which time the nurse will assess for blood return and the ability to infuse. If the problem still exists, this process can be repeated at the same dose (1mg/1ml).

## TPA Administration for Intra-abdominal/Chest tube catheters

TPA can be administered in non-vascular catheters. Chest tubes and abdominal drains can become occluded after insertion. Administration of fibrinolytics can re-establish patency. At attending discretion, TPA can be ordered in ratio of 1mg TPA to 10ml Normal Saline. For instance, in an adult size adolescent an order of 4mg TPA in 40ml NS is appropriate and can be administered through a chest tube or catheter. Due to limits in volume, small patients might require small doses 2mg/20ml NS or 1mg/10ml syringes. Using aseptic technique the syringe should be attached appropriately to luerlock or the available port of 3-way stopcock. Administration/injection of the TPA should continue until patient endorses pain or until full volume is used. Upon completion of instillation the catheter must be clamped for 60-90 minutes. Nursing must be made aware of this as they will need to record the total output of infusion as well as the amount that drains out at the conclusion of the 60-90minutes.

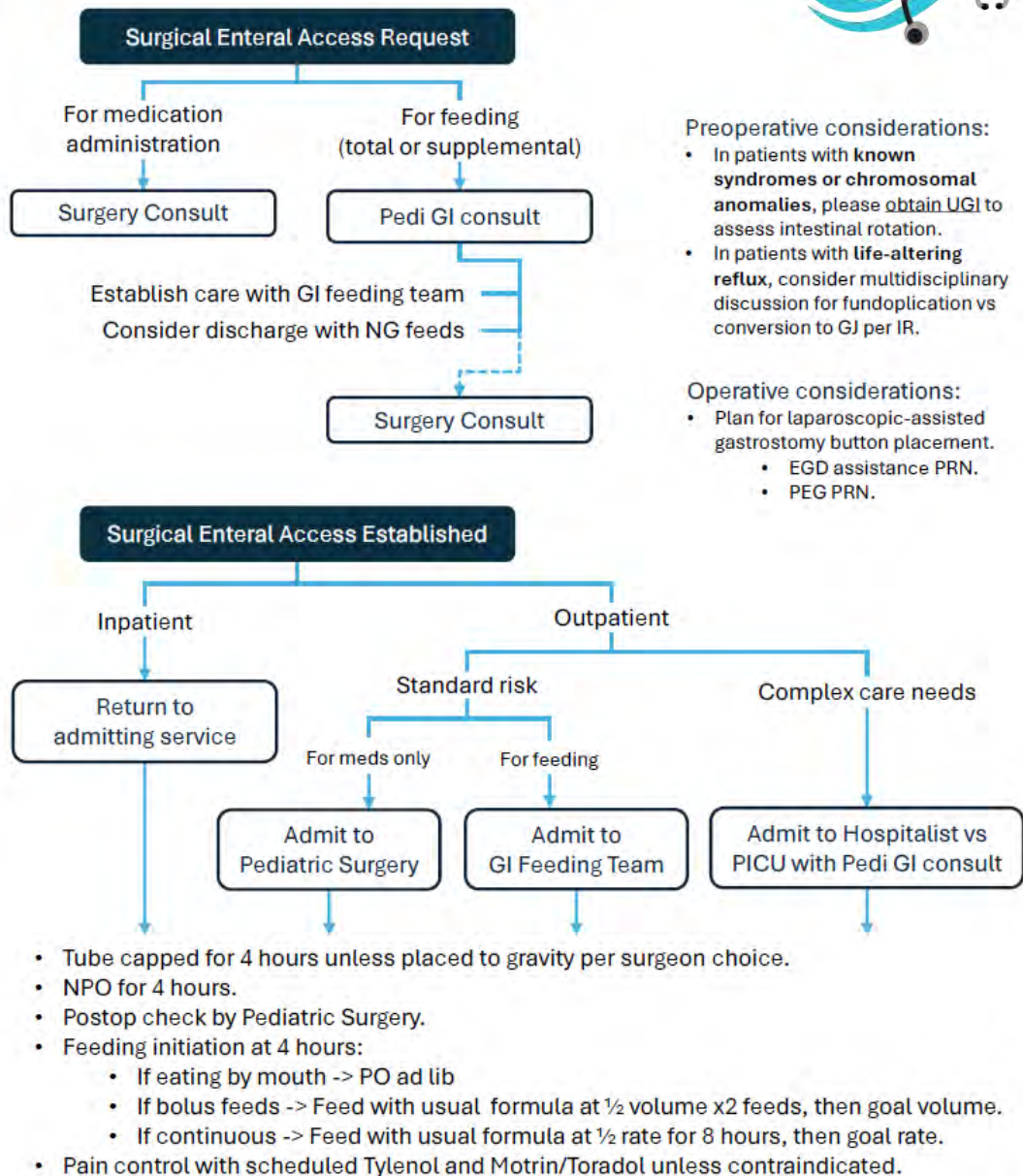
## Central Line Infections

Patients may need tunneled lines for chronic TPN at home. When such a patient presents with fever, the line is assumed guilty until proven innocent. The flow diagram below describes the short gut service's approach to the evaluation and treatment of suspected line infections in children on home TPN.





## Surgical Enteral Access Guidelines



## NON-OPERATIVE MANAGEMENT OF ACUTE APPENDICITIS

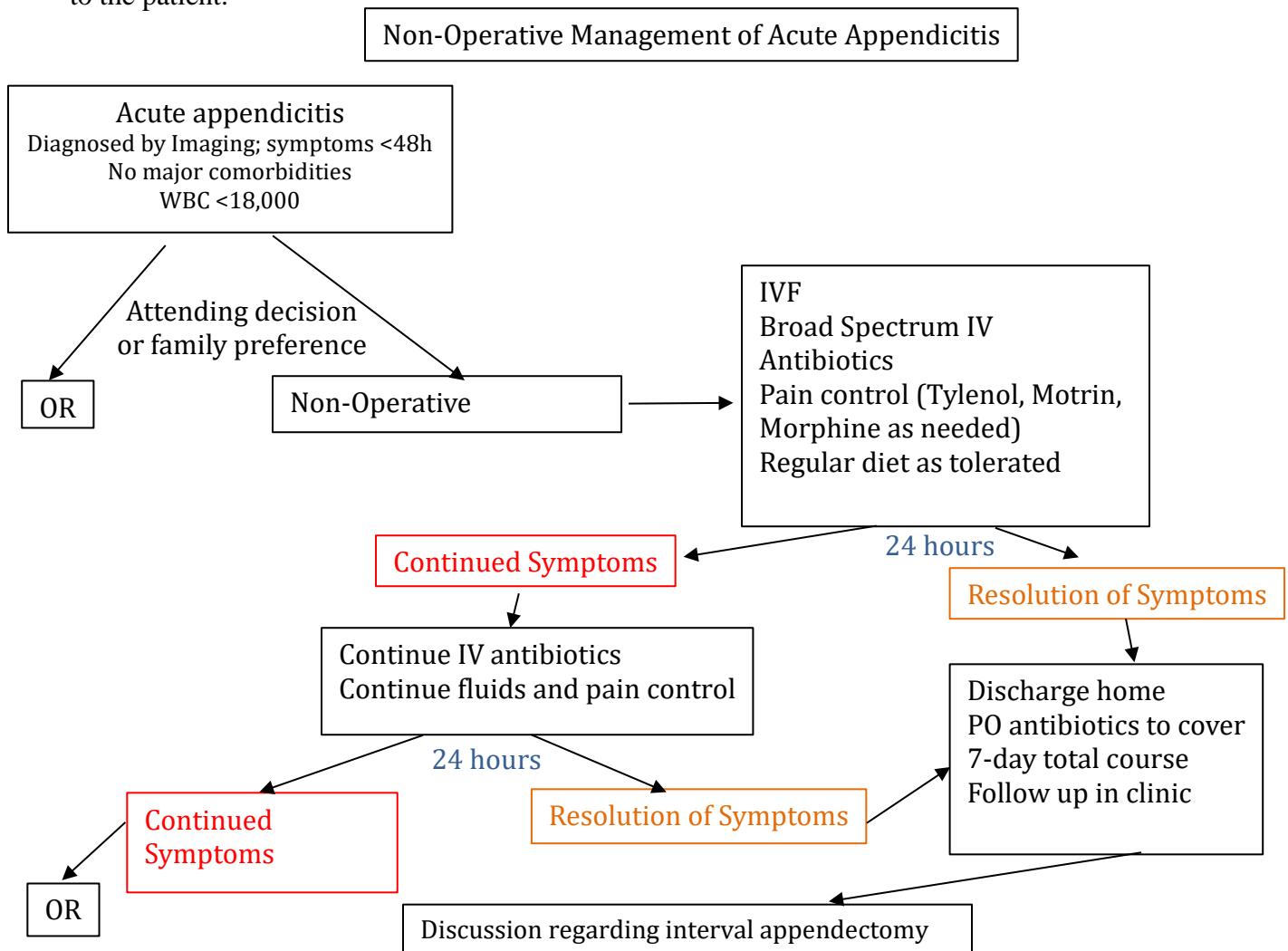
The Division of Pediatric Surgery currently has two care pathways for treatment of appendicitis: non-operative management of acute appendicitis, and advanced appendicitis.

### Non operative management of acute appendicitis:

The treatment of acute appendicitis non-operatively is controversial. There is evidence that it can be used effectively, but there is a high recurrence rate of appendicitis within one year of treatment (as high as 30%). The option to treat non-operatively **MUST** be discussed with your attending before presenting it as a potential treatment option for patients. This pathway may be an option for patients who are COVID positive or have some other co-morbidity that would increase the risk of general anesthesia.

The broad-spectrum antibiotic usually chosen is Zosyn. Ciprofloxacin and flagyl may be an option in penicillin-allergic patients.

By the current pathway, patients are considered a “failure” of non-operative management if they have persistent unresolving abdominal pain, inability to tolerate PO, or fever at 48 hours. At this point surgery would be considered. Again, discuss with your attending before presenting the plan to the patient.

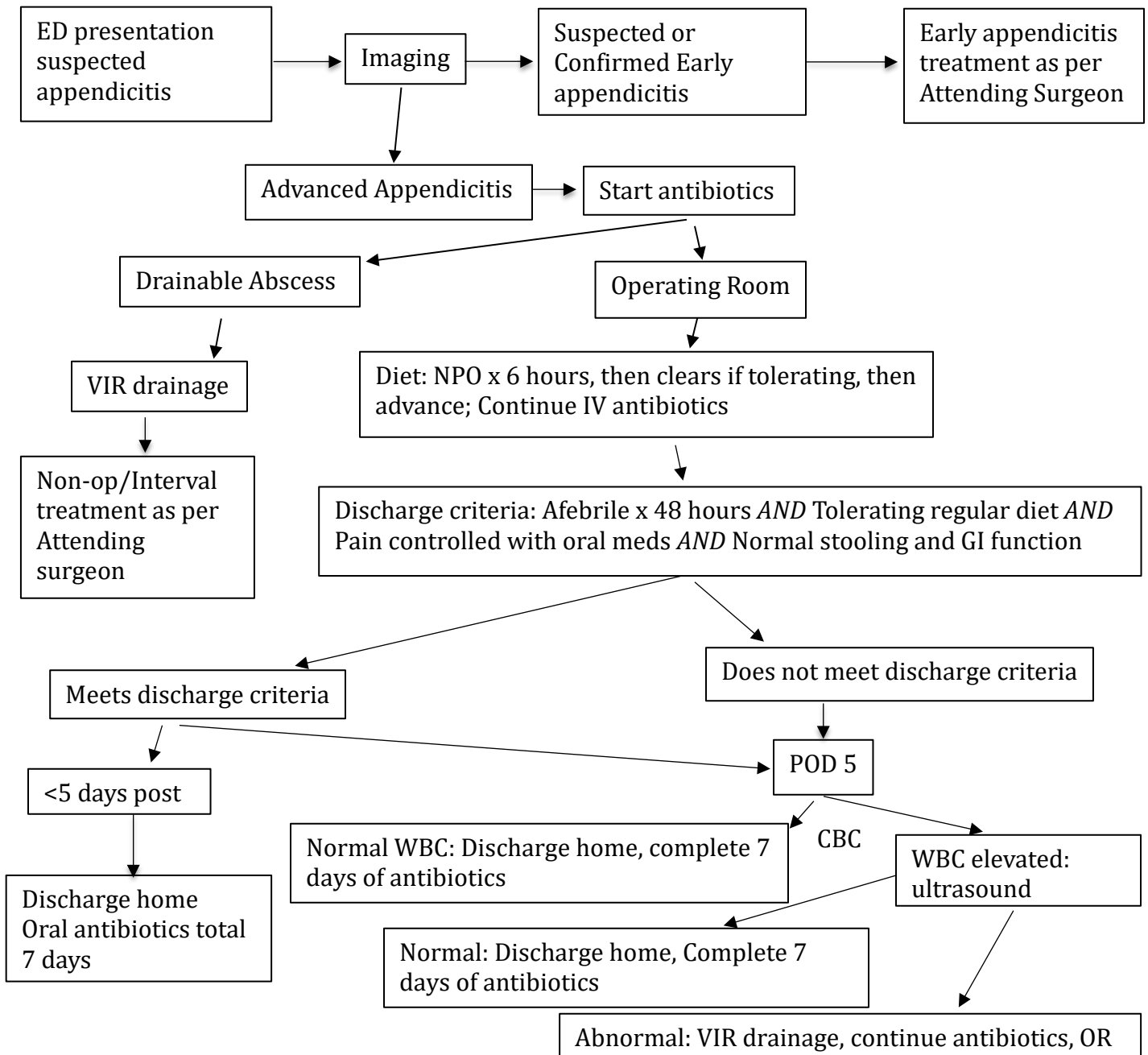


### Advanced Appendicitis Pathway:

“Advanced appendicitis” refers to any form of appendicitis beyond acute – gangrenous, suppurative, and perforated. This pathway addresses the operative management of these forms of appendicitis. We currently do not have pathways for non-operative management of advanced appendicitis or for management of appendicitis that presents with a well-defined abscess. Please discuss these two situations with your attending.

The most common antibiotic choice is Zosyn, with Ciprofloxacin/Metronidazole for penicillin allergy.

Definitions: *early appendicitis* – acute, non-suppurative by radiology; *advanced* – gangrenous, purulent, perforated by radiology



## ILEOCOLIC INTUSSUSCEPTION

Definition: Intussusception occurs when a part of the intestine folds into itself like a telescope, with one segment slipping inside another segment.

Presenting Symptoms/Signs: Patients usually present between 3 months and 4 years of age. They will experience intermittent colicky abdominal pain occurring every 15-20 min with drawing up of the legs, abdominal distention, vomiting or passage of bloody and/or jelly-like stool. These symptoms may have been present for only a few hours or for several days. Intussusception may be seen more frequently in cooler seasons when viruses are prevalent. With later presentations, a child maybe dehydrated, septic and/or lethargic.

Cause: Due to alterations in bowel motility, although a lead point may be present, especially in older children. Lead points include Meckels diverticulum and polyps. Intussusceptions cause intraluminal obstruction, proximal distention, potential vascular compromise to blood supply, dehydration, and sepsis.

Evaluation/Management: The Pediatric Surgery team will be consulted by the ER to help with suspected bowel obstruction (colicky pain followed by distension), abdominal pain/vomiting, or lower GI bleed. The child will likely have had blood work, an AXR and a confirmatory ultrasound of abdomen demonstrating intussusception.

### Evaluation and Management

- Start with the ABCs, followed by assessing the overall wellness of the child
- Obtain your own history from the caregiver including the length of symptoms
- Examine the child to assess the severity of dehydration, presence of abdominal mass, and any evidence of peritoneal irritation
- Resuscitate the child with appropriate fluids depending on your evaluation of the degree of dehydration and monitor urine output
- Review the ultrasound to confirm the diagnosis. This is usually the definitive study for diagnosis of this disease.
- Some surgeons may consider starting a prophylactic antibiotics on making the diagnosis while waiting for treatment; check with your attending for guidance on this issue**

### Treatment

Either radiologic or surgical.

- If the child is hypo-responsive with a prolonged history of symptoms (> 3-4 days) or has signs of peritonitis, a surgical option may be considered early.
- In a stable patient, an air enema can be used to reduce the intussusceptum from intussusciens.
- The reduction is performed by a radiologist and the resolution of intussusception
- is confirmed by reflux of air into the proximal portion of ileum or intestine.
- **Patients need an IV prior to arriving in radiology.**
- It is important for **the pediatric surgical team to be present during air enema reduction.**
- Air enema reduction may cause bowel perforation and even cardiorespiratory arrest due to rapid insufflation of abdomen and potential compromise of respiration and circulation. In addition to rapidly arranging for surgical intervention, one should be prepared to **decompress** the abdomen if tension pneumoperitoneum is present by careful insertion of a #14 gauze angio-catheter through the abdominal wall

- Depending on how far the intussusceptum has advanced into colon, the radiologist
- may elect to reduce in multiple attempts (usually 3-4 times) over several hours
- depending on their progress.
- The general incidence of recurrence following air enema reduction is ~10 %.
- Patients are admitted for observation overnight following the initial reduction (pathway for ED discharge is currently being drafted)
- If symptoms recur, the air enema will be repeated.
- One should consider a lead point as a cause of intussusception if there are multiple
- recurrences or failure of reduction

## Surgical indications for intussusception

In addition to peritoneal signs with prolonged presenting history, include failure to reduce and multiple recurrences with the confirmation of a lead point as a cause of intussusception. Consent for laparoscopy or laparotomy, possible reduction of intussusception, possible bowel resection, possible stoma should be obtained

The above information primarily describes the evaluation and treatment of ileocolic intussusception. Pediatric Surgery may occasionally be consulted for small bowel-small bowel intussusception. This entity can often be seen on ultrasound when a patient has gastroenteritis or other conditions increasing bowel motility. Most often it is not a cause of symptoms and will self-resolve. Occasionally, it may need treatment depending on the duration of persistence of the intussusception on imaging, the length of the intussusception, signs of bowel obstruction proximal to the intussusception, or presence of a lead point. Conditions such as Peutz-Jeghers or Henoch-Schonlein Purpura may increase the likelihood of a lead point and need for treatment. Patients should be individually evaluated and discussed with your senior and attending.

### Reference:

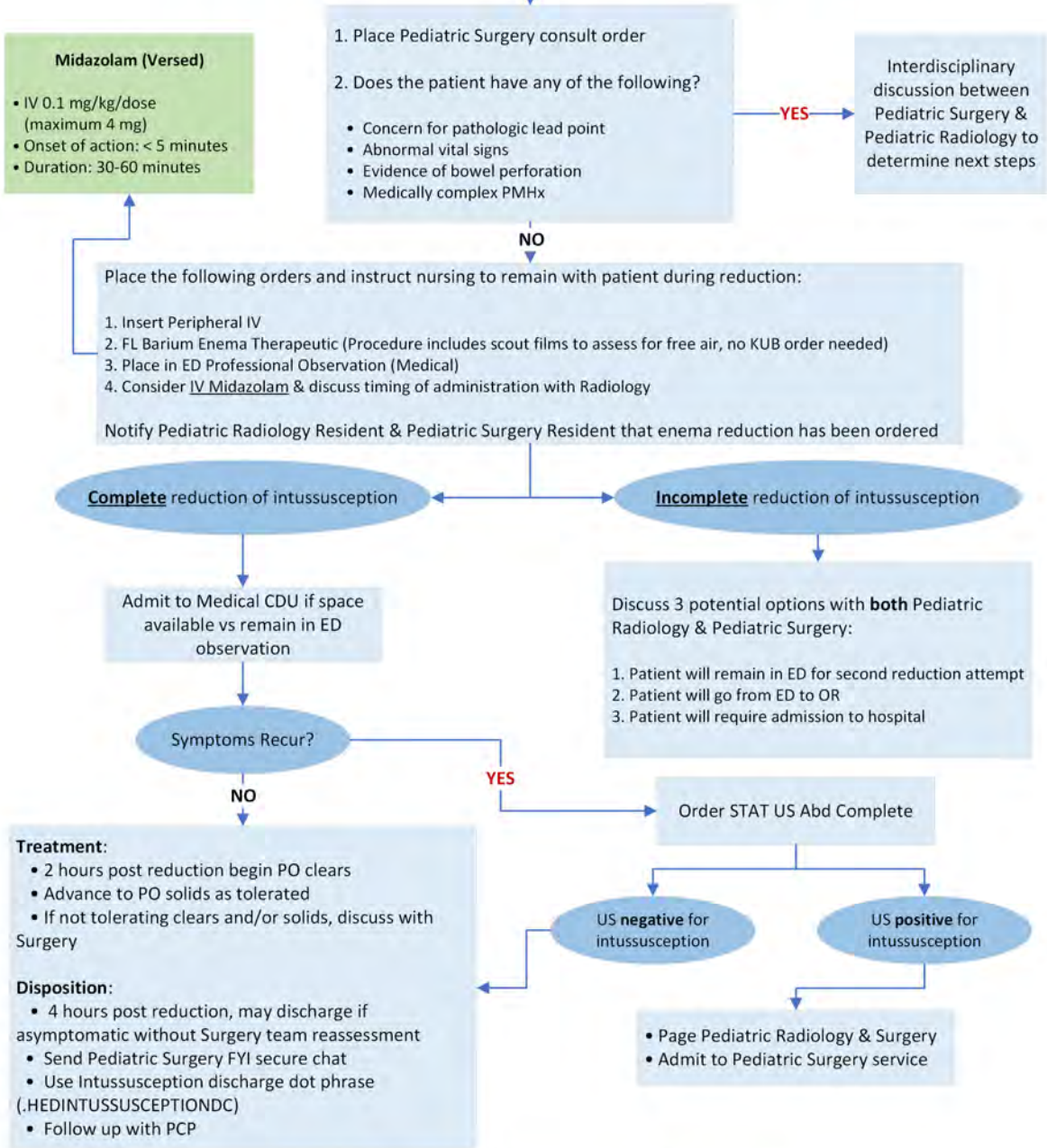
1. Diagnosis and treatment of Intussusception- A surgical condition Mark M. Ravitch Pediatrics 1966;38:122
2. Pediatrics International (2012) 54, 948–958, e35–e42



# Intussusception Guideline

Last Updated: 9/2024

## Ileocolic Intussusception on Diagnostic Imaging





Last Updated: 9/2024

# Intussusception Guideline

## Discharge Instructions

Type **.HEDINTUSSUSCEPTIONDC** into the discharge instruction for the following information:

Your child was diagnosed with and treated for **intussusception**.

### What is intussusception?

It is a condition that can cause abdominal pain. It occurs when one part of the intestine slides into another part of the intestine. When this happens, it causes a blockage. When a blockage occurs, air, fluid, and/or food can get stuck which can cause symptoms.

### What are the symptoms of intussusception?

Symptoms include but are not limited to sudden abdominal pain which can get better then come back, throwing up, bloody poops, fits of crying, and being very sleepy or hard to wake.

### What causes intussusception?

The cause is usually not known. It can happen after a child has a viral illness.

### Can intussusception come back?

Yes. It is most likely to recur in the first day or two after treatment. The symptoms of a second intussusception are the same as the first time.

### What happens when my child goes home?

We recommend you follow up with your Pediatrician for a repeat examination in 1-2 days from discharge.

### When should I return to the Emergency Department?

Return to the Emergency Department if your child has blood in their poop that is not going away, is very tired or hard to wake up, or has a swollen abdomen.

### Who should I contact if I have questions?

You can always call your child's Pediatrician or contact the Pediatric Surgery team that took care of you in the Emergency Department. Their contact information is listed below.

Center for Pediatric Surgery at Brown Surgical Associates  
2 Dudley Street Suite 190 02905-3236  
Phone: 401-421-1939

## SKIN AND SOFT TISSUE INFECTIONS

Both cellulitis and abscess are common skin and soft tissue infections in children. Cellulitis typically appears as erythema, edema and warmth after bacteria has violated the skin barrier. An abscess is a collection of fluid, pus and/or blood within the dermis or subcutaneous space. Both may occur in otherwise healthy children with no known predisposing factors. The most common cause of cellulitis is Streptococci, while the most common pathogen associated with abscess is Staphylococcus aureus.

### Patient evaluation

- Identify risk factors for MRSA (family or personal history, family member in healthcare etc.)
- Identify underlying medical conditions (malignancy, immunocompromised state)
- Ask about a history of prior abscesses
- Evaluate for recent trauma (including animal or insect bites), recent rashes, use of antibiotics (sometimes prescribed by a Pediatrician prior to Surgery evaluation)
- Obtain travel history
- Ask about allergies to medications

### Physical Examination

- Temperature, heart rate, blood pressure
- Evaluate for erythema, Induration, fluctuance, presence of pustule
- Ask if the area has spontaneously drained or been previously drained
- Note: ultrasound is not necessary for diagnosis!

### Treatment

#### Incision and Drainage

- Can be performed in the Emergency Department, Pediatric Sedation, Operating Room
- If performed in ED, the need for sedation is based on patient age, medical history, associated medical disorders, location/size of abscess and timing of last PO
- Discuss sedation options with ED provider. These may include IN Fentanyl, IN Versed, IV Morphine, IV Versed, IV Ketamine
- Discuss plan for incision and drainage with patient and family and obtain consent for the procedure. If being done under moderate sedation in the Emergency Department or Pediatric Sedation consent is not needed because the procedural consent is included as part of the sedation consent
- Document consent and procedure in a Procedure Note
- Supplies to gather:
  - Local Anesthetic: Lidocaine
  - #11 blade
  - Chlorhexadine prep OR Betadine
  - Towels
  - Gauze

- Normal Saline 10 cc flush
- Vessel loop OR penrose drain OR plain/Iodoform packing \*\*discuss choice with Attending
- Culture swab
- Kelly OR Jake OR Mosquito
- Suture material if needed
- Dressing supplies (gauze, Tegaderm, tape, Hypafix etc.)
- Discuss details of the procedure with Senior On-Call prior to performing I&D. This will guide decision making in terms of drain placed and wound care to follow.

### Antibiotic Therapy

- Indications: Cellulitis and induration with systemic signs and no indication for I&D OR persistent cellulitis and systemic signs after I&D
- Nafcillin 150 mg/kg/day divided Q6H AND Clindamycin 25-40 mg/kg/day divided Q8H
- PO options if patient is to be discharged:
  1. Clindamycin 30-40 mg/kg/day divided Q8H
  2. Keflex 25-50 mg/kg/day divided Q6H
  3. Bactrim 8-12 mg/kg/day divided Q12H

### Wound Care

- Warm Compresses: usually used if an area of cellulitis is not yet “ripe” enough for drainage and the patient is on antibiotics. May encourage “pointing” of the infection which will allow drainage
- Sitz baths: often used after pilonidal abscesses or perineal/labial abscesses
- VNA care: sometimes needed, depending on comfort level and resources of family
- If a packing is left, it should be removed within 48 hours by family or VNA
- If a drain is left, it should be removed in Pediatric Surgery Clinic 3-5 days post-operatively
- Tylenol and/or Motrin for pain control with wound/dressing care

## PILONIDAL DISEASE

Pilonidal disease refers to pits, abscesses, sinuses or pseudocysts of the natal cleft. The disease varies in presentation. The exact cause of pilonidal disease is unknown, but hair plays a role. The hair involved is most likely “lose hair” that, as a result of friction within the intergluteal cleft, breaks off and falls into an open pit. Filled with hair and debris, these pits serve as a site of incubation for bacteria. Pilonidal disease is most common in teenage males. Risk factors for the disease include increased weight, sedentary lifestyle, a deep natal cleft, and excessive hair growth.

### Patient presentation with acute disease

#### History and symptoms

- Pain within intergluteal cleft
- Swelling or redness
- Bloody, mucoid or purulent drainage
- Fever

#### Physical Examination

- Findings are superior to, lateral to or within natal cleft
- Erythema
- Tenderness
- Fluctuant Mass
- Midline Pits
- Sinus Tracts
- Granulation Tissue
- Bloody, mucoid or purulent drainage
- Excessive hair growth

### Treatment

#### Incision and Drainage

- Performed in Emergency Department, Pediatric Sedation, or Operating Room
- If performed in ED: assess the need for sedation based on patient age, medical history, associated medical disorders, and timing of last PO
- Discuss sedation options with ED provider. These may include IN Fentanyl, IN Versed, IV Morphine, IV Versed, IV Ketamine
- Discuss plan for incision and drainage with patient and family and obtain consent for the procedure. If being done under moderate sedation in the Emergency Department or Pediatric Sedation, consent is not needed because the procedural consent is included as part of the sedation consent

- Document procedure and consent in a Procedure Note
- Incise over point of maximum fluctuance
- Incise off midline
- Supplies Needed:
  - Local Anesthetic: Lidocaine
  - Chlorhexadine prep OR Betadine
  - #11 blade
  - Culture swab
  - Kelly OR Jake OR Mosquito
  - Tweezers
  - Saline Flush
  - Razor OR Clippers
  - Plain Packing versus Vessel Loop versus Penrose Drain with stitch
  - Gauze
  - Tape
  - Tegaderm OR Hypafix

## Antibiotics

- Consider only if cellulitis and tenderness with no evidence of fluctuant mass OR if extensive cellulitis remains following I&D
- Oral Ciprofloxacin, Flagyl, Augmentin, Clindamycin, Bactrim

## Wound Care

- Provide extensive education to family regarding wound care (review with senior and attending)
- If packing is present, removal to be performed within 48 hours
- If drain or vessel loop is present, removal to be within 3-5 days in Pediatric Surgery Clinic
- Sitz Baths, Showering, dressing changes may be part of wound care
- Hair removal (once acute wound is healed): depilatory cream, laser hair removal
- Tylenol and/or Motrin for pain control with wound/dressing care

## Definitive treatments

### Pit Picking

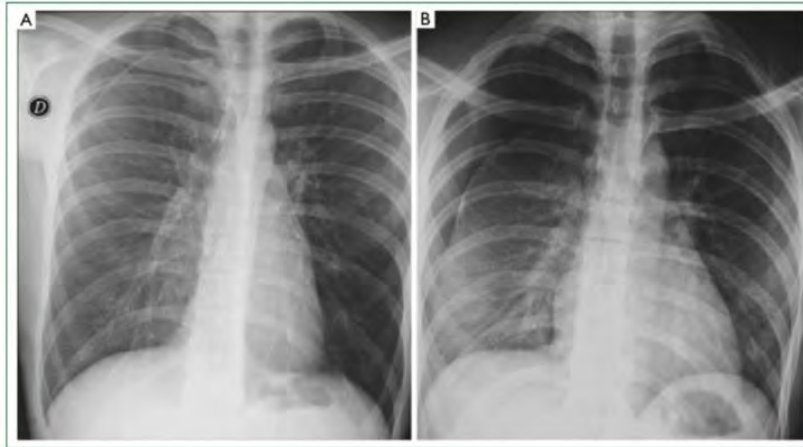
- Minimally invasive
- Performed in Operating Room
- Discharged to home following procedure
- Wound care dictated by operating Surgeon

### Excision

- Options include marsupialization, wide excision with secondary healing, wide excision with primary midline closure and excision with off-midline closure

## PNEUMOTHORAX

Definition: Pneumothorax refers to the presence of air in the pleural space between the lung and the chest wall. “Primary spontaneous” pneumothorax occurs without any precipitating traumatic or iatrogenic mechanism. When there is a known cause, the pneumothorax is “secondary” to the precipitating event.



### Cause

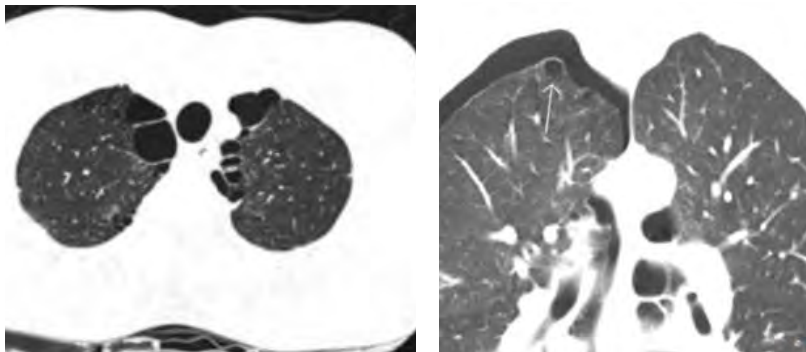
Most commonly, emphysematous blebs, asthma, and tobacco have been attributed. It is commonly thought that young males with Marfan syndrome, a common inherited disorder of connective tissue, are prone to spontaneous pneumothorax. If younger children present with spontaneous pneumothorax or recurrent pneumothorax, uncommon underlying diagnoses including CPAM and pleuropulmonary blastoma may need to be considered.

### Evaluation/Management

The pediatric surgical team will be asked to consult in ER when spontaneous pneumothorax is suspected (pleuritic chest pain with or without shortness of breath). The child will likely have had blood work and CXR (sometimes with insp/exp views).

1. Always start with the ABCs, followed by getting a general feel for acuity of the shortness of breath or severity of dyspnea including color, resp rate and nasal flare etc
2. Obtain your own history from the patient or caregiver including the length of symptoms
3. Examine the child to confirm your assessment for the severity of RR, SOB, saturation and any evidence of tracheal shift or tension pneumothorax:
4. Assess and resuscitate the child with appropriate respiratory support while conducting your history and examination
5. Depending on the acuity and severity of pneumothorax, the surgical team will place a pigtail chest catheter or tube urgently or emergently with local anesthesia and possibly mild sedation. The tube should be **STABLY** secured to the chest wall. A follow up chest X-ray should be obtained immediately to check the position of the chest tube and expansion of the lung while the chest tube is under 10-20 cm suction
6. Make sure that the patient is comfortable with the chest tube in situ, and consider incentive spirometry to encourage full expansion of the lung:

7. Although there may be a variance in opinion, some surgeons prefer to get a CT scan of chest during the first visit to document suspected causes such as blebs in the apices of upper and lower lobes
8. Recurrent pneumothorax is the most common complication after an initial episode and has been documented in 30-50% of children. Primary spontaneous pneumothorax recurrence rates are highest (22.4% to 36.8%) in children 13 to 18 years of age, with almost half developing ipsilateral recurrence and 14% developing contralateral recurrence. Most surgeons would offer surgical treatment on recurrence while most primary presentations will be treated expectantly with chest tube.



### Operative Intervention

Surgery usually consists of Video Assisted Thoracoscopy with resection of any blebs and mechanical or chemical pleurodesis. Post operatively, the chest tube will be kept to suction for a period of time (check with your attending for duration) before removal in order to keep the lung expanded and to promote adhesion formation. While pneumothorax can recur after surgery, the pleurodesis helps to prevent complete collapse of the lung and to mitigate accompanying symptoms.



### References:

- a. Management of spontaneous pneumothorax. An American College of Chest Physicians Delphi Consensus Statement. *Chest* 2001;119: 590-602
- b. Management of Spontaneous Pneumothorax. *Chest*. 2015;148(4\_MeetingAbstracts):435A. doi:10.1378/chest.2280584

## COLORECTAL BUNDLE

Colorectal procedures have a high risk of surgical site infection. In adults, bundles can reduce this risk. The Pediatric Surgery Service has developed a Colorectal Bundle which is tailored to the needs of its pediatric patients; this differs slightly from the one developed for adult patients.

### Bundle elements:

- Preoperative Bath night before and/or morning of surgery
- Can be: Bathing with soap and water; bathing with chlorhexidine-containing solution; wiping with a chlorhexidine-impregnated cloth; non-medicated bath cloths
- Hair removal: Age-appropriate hair removal with clippers prior to draping, otherwise omit
- Glucose testing: For patients who are known diabetics or receiving preoperative therapies that increase the risk of hyperglycemia (steroids), glucose testing is performed immediately after induction of anesthesia
- Oral antibiotic prep: Oral antibiotic prep the day before surgery for elective procedures (Neomycin 15 mg/kg/dose, maximum 1000 mg/dose, and Metronidazole 15 mg/kg/dose, maximum 1000 mg/dose, at 7 pm and 11 PM)
- Ancef/flagyl dosed within 60 minutes prior to incision, with substitutions for patients with allergies (Vancomycin and fluoroquinolones administered 0-120 minutes prior to incision)
- Ancef re-dosed if case longer than 3 hours, or appropriate re-dosing for substitutions
- Temperature: Patient temperature  $\geq 36.0$  C from incision to wound closure (monitored with pediatric-appropriate probes)
- Change of gloves and gowns before closing
- Use of dedicated age appropriate “clean closure” tray for closing

### Notes:

- Does not include patients < 1 month of age
- Ask attending whether the Bundle applies to a patient’s operation
- Depending on the case, other preop measures may include mechanical bowel prep, enemas, or clears for a defined period before surgery. Please review these details with the attending.

## OVARIAN TORSION

**Definition:** Twisting of the ovarian vascular pedicle, sometimes involving the fallopian tube, which results in:

**Lymphatic and venous obstruction → edema → arterial occlusion → ischemia and necrosis of ovary**

### Risk factors

- **Ovarian enlargement** due to a cyst or other mass
- 51-85% of torsions associated with a cyst or mass
- Cyst > 5 cm increases the risk of torsion
- **Elevated hormone levels** (ovulation induction, PCOS, maternal exposure in the neonate)
- **Abnormally long fallopian tube, mesosalpinx, mesovarium**

### Classic Presentation

- **Sudden severe** unilateral abdominal or pelvic pain
- Associated nausea, vomiting, dysuria - can be synchronous with onset of pain
- Exam: tenderness, possible unilateral mass on palpation

### Differential diagnosis

- Premenarchal
- Appendicitis, nephrolithiasis, mesenteric adenitis, intussusception, UTI, gastroenteritis
- Adolescent
- Pelvic inflammatory disease, tubo-ovarian abscess, ectopic pregnancy, ovarian cyst, hemorrhagic cyst, ovarian mass, Mittelschmerz, UTI, gastroenteritis

### Studies

#### Labs

- Pregnancy test if post-pubescent
- If mass is suspected:  $\alpha$ FP,  $\beta$ -HCG, inhibins A & B, CA-125, CEA, CA 19-9, LDH
- Imaging

#### Ultrasound

- Doppler for blood flow: can be unreliable as arterial flow may be present in torsed ovaries and may be absent in normal ovaries of a young patient
- Twisted vascular pedicle – harder to see in young children
- Atypical location – midline or on contralateral site from origin
- Adnexal mass or cyst
- Morphology important
- Unilateral ovarian enlargement due to edema or hemorrhage
- Compare with established normal volumes for age
- $\geq 3x$  volume of contralateral side – concerning for torsion

- Complex or cystic structure
- Follicles only on periphery or absent
- Follicular enlargement with fluid-debris levels: indicate edema and hemorrhage
- Drawbacks: Ovaries are smaller and more difficult to identify in young pediatric patient than they are in adults; Pediatric ovaries may be displaced into abdomen rather than in pelvis

## MRI

- Findings
- Ovarian enlargement
- Central hemorrhage and edema with peripheral follicles
- Poor enhancement suggesting infarction or necrosis
- Contrast enhancement can detect swirling of twisted pedicle
- Disadvantages
- Cost, time for exam

## Treatment

### Surgery required

- Laparoscopy for detorsion and ovarian preservation; can perform open if necessary
- Cystectomy can be considered if cyst present; oophorectomy if tumor confirmed
- Attempt to preserve ovary if possible
- Black or necrotic ovaries can still be treated with detorsion and possible preservation
- Folliculogenesis has been well documented following ovarian detorsion
- Suspected tumor
- 1.8% risk of malignancy in masses found in torsion in pediatric patients; increased risk if mass >8cm
- Interval evaluation can be considered for torsed ovaries if mass is suspected but there are no overt signs of malignancy
- Detorse, leave mass in place, check tumor marker results, interval surgery

## Recurrence

- Due to anatomic predisposition: laxity of utero-ovarian ligaments, long fallopian tube
- Risk range 5-18%
- Oophoropexy: plication of adnexal ligaments or fixation of adnexa using non-absorbable suture to pelvic sidewall
- Concern about impact on fallopian development or fertility
- Evidence that recurrence can still occur after pexy; no evidence to support pexy at first episode of torsion

## Follow-up:

Consider imaging around 3 months to document presence of ovarian follicles, sooner if concern for neoplasm. Preferably, obtain follow-up imaging at a *different* time in the patient's cycle: e.g. 6, 10, 14 weeks later.

# OVARIAN CRYOPRESERVATION PROGRAM

## Who is this program for?

Pediatric oncology patients who are pre-pubertal or who cannot undergo traditional egg harvest AND are at high risk for gonadal dysfunction/infertility from their planned therapy.

## What does this program do?

Safe unilateral oophorectomy for cryopreservation

## When do these patients undergo surgery?

- Prior to initiation of cancer directed therapy. Ovarian harvest generally will be paired with other surgical procedure (ie central line placement)
- Procedures will need to be first case of the day Monday-Thursday in order to get the sample to Boston by 12pm

## Why are we doing these procedures?

- To allow patients the opportunity to have children using their own eggs in the future
- Onco-fertility is an important part of the future of cancer therapy as we cure more childhood cancers.

## Onco-Fertility Team

Pediatric Oncology: Roma Bhuta, DO, MPH

BrownHealth Laboratory: Dariusz Stachurski, MD

RIH send out lab: Carole Kavanagh, Debra Napert

Pediatric Surgery: Elizabeth Renaud, MD

Adult Gyn Onc: Don Dizon, MD

Reproductive Endocrinology: May-Tal Saurbruncutle, MD

IVF Lab Technical Director: Jay Patel, Brigham and Women's Hospital

BWH onco-fertility nurse/coordinator: Patricia Kennedy, RN

## Protocol

### Eligibility

- Female patients unable to pursue egg harvest due to:
  - Prepubertal age
  - Unable to wait the necessary ~2 week time frame prior to starting therapy for malignancy
  - Patients who need intensified therapy that is highly gonadotoxic due to relapsed or refractory disease
  - Any age (Boston has done as young as 4 months)
  - Female patients at high risk for infertility, low risk of ovarian involvement of disease
- XRT
  - Whole abdomen or pelvic XRT (>15-20 Gy)
- TBI
- Alkylating intensive chemotherapy

- Solid tumors
- Pre-HSCT (when patient in remission)
- Conditioning regimen with alkylator/TBI
- No contraindication for oophorectomy

### Exclusion criteria

- Surgical contraindication for oophorectomy
- Coagulopathy, platelet count, hostile abdomen from previous procedures, solitary ovary
- Known involvement of ovary with malignancy
- New hematologic malignancy diagnosis (leukemia)
- Patients who will receive treatment that is overall low risk of gonadal dysfunction (determined by oncology team)

Consents (to be obtained by Oncology Team; surgical consent is obtained by surgical team)

- Partners PHI release form to allow Brigham and Women's Hospital to release information to storage facility (Patient signs)
- Rhode Island Hospital PHI release for RIH to BWH (RIH physician)
- BWH Ovarian Tissue Consent (Patricia Kennedy, RN)
- BWH Egg Freezing consent (Patricia Kennedy, RN)
- BWH Tissue Release Form to release to storage facility (Patricia Kennedy, RN)
- Financial Consent for BWH processing (Patient signs)
- NE Cryogenic Center Consent Packet (if applicable, patient signs)
- RIH consent for local storage and transport to BWH (RIH physician)

### **Multidisciplinary Contacts once patient found to be eligible and interested in harvest (to be done by Oncology team)**

- Notify embryologist at BWH
- Jay Patel: [jpatel35@bwh.harvard.edu](mailto:jpatel35@bwh.harvard.edu)
- Cassy Thomas: [crthomas1@bwh.harvard.edu](mailto:crthomas1@bwh.harvard.edu)
- Berhan Bogale: [bbogale@bwh.harvard.edu](mailto:bbogale@bwh.harvard.edu)
- Tina Cappello: [tcappello@bwh.harvard.edu](mailto:tcappello@bwh.harvard.edu)

### Financial

- Oophorectomy
- Insurance
- Transport from RIH to BWH
- No cost to patient, price incurred by hospital
- Tissue processing at BWH
- Out of pocket; ~\$1090
- Email billing at BWH to charge patient. Christina Francis: [cfrancis4@bwh.harvard.edu](mailto:cfrancis4@bwh.harvard.edu)
- Storage
- NE Cryogenic Center

- ~\$475/year
- If eligible for Verna's purse\*
- Reprotect (sends to MN or FL)
- ~\$90/year
- Required infectious disease testing for storage at cheaper cost
- Transport to storage facility
- NE Cryogenic Center ~\$250
- Incorporated into NECC charges for storage (includes transport + 1<sup>st</sup> year storage)
- Reprotect ~\$350

\*Eligible for Verna's purse if household income <\$100,000 (need proof via recent tax form). If family interested, family needs to apply online + separately also register with Reprotect

[ReproTech Financial Assistance](https://www.reprotect.com/forms/)

<https://www.reprotect.com/forms/>

## Scheduling

- Ideally paired with other surgical procedure (ie central line placement)
- Oophorectomy must be done Mon-Wed as sample can only be processed at BWH Mon-Thursday prior to 12pm
- Aim for first case in the morning for same day delivery to BWH
- Second option: Plan for afternoon case. Tissue must be at 4C or on wet ice <20 hours total
- When an ovarian cryopreservation case is anticipated, the surgeon should secure chat Brian Mullahey and Courtney Bouchard from the booking office. At least 2 days of advanced notice are preferred in order to facilitate timing and coordination of scheduling.
- Send out Lab to be notified in advance of surgical procedure (4-6109)

## Surgical procedure

- Consent
- Should include a list of complications, including bleeding, infection, injury to intraabdominal structures, injury to fallopian tube, hernia formation
- Should explain that complications may delay the start of chemotherapy, depending on type and severity
- Side should not be stated. Usually, the right ovary will be used, but ovaries will be assessed intraoperatively and the larger of the two may be used
- Should emphasize that this is an elective procedure that will not be performed if it may delay curative therapy or if other safety criteria are not met
- Laparoscopic unilateral oophorectomy
- Day of surgery: surgery will not be performed if Hb <10, plt < 50, ANC < 500
- May discuss the use of GCSF with oncology team
- Ancef for antibiotic prophylaxis
- A foley catheter will be placed for the duration of the case to assist in visualization for ovarian harvest; removed before wakeup

- Laparoscopic entry at umbilicus
- Evaluate for presence of 2 ovaries (solitary ovary is a contraindication to procedure)
- Ovary for harvest: usually the right ovary (do not need to contend with sigmoid colon) or larger ovary if there is a significant size discrepancy
- Delicate handling of ovary: The ovary should not be directly handled with instruments. It can be manipulated by holding the mesovarium or lifting the fallopian tube
- Fallopian tube should not be harvested and should not have blood supply harmed during procedure; can be manipulated during procedure for exposure to avoid handling trauma to ovary
- Secure ovarian pedicle with ligasure, harmonic, or cool seal; seal should be twice burned with ligasure or cool seal before cutting
- Place in endocatch bag for retrieval
- Umbilical incision should be enlarged to easily extract ovary without damaging or compressing the specimen
- Ovary placed in transport media immediately after extraction (NO FORMALIN)
- Tissue is placed in a 50ml conical tube or specimen cup filled with MEM media (HEPES buffered media, preferred manufacturer Sigma; see picture below) and tube/container placed on or slightly submerged in wet ice. No formalin
- OR will call send-out department and a lab member will come pick up the specimen directly from OR. No hospital courier

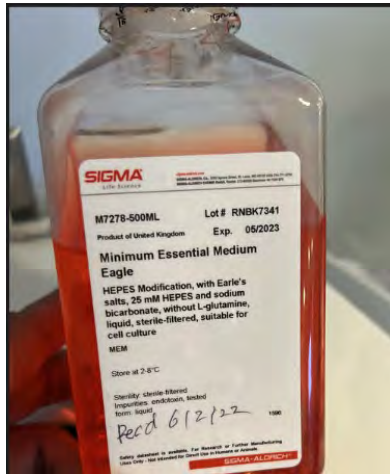
## Send out Lab

- Will package tissue container and STAT courier will be called for direct transit to BWH embryology laboratory
  - Sample packaged in small styrofoam cooler. Package does not need to be marked; just ensure it is secure.
  - Sample to be kept on wet ice throughout duration of transport
- Brigham & Women's Hospital
- IVF Embryology
- Francis Street |Tower5C| Boston, MA 02115.
- Phone: 617-732-5157
- Send out lab will call BWH embryology lab as soon as courier has picked up specimen
  - Jay Patel, cell: 339-970-4245
  - BWH lab: 617-732-5157
- Ideally same day delivery of ovarian tissue to BWH
  - Delivered to BWH Mon-Thurs by 12pm
  - Transport tissue on wet ice
- Next day delivery to BWH (if same day not possible)
  - Store tissue in MEM media at 4C overnight and transport on wet ice morning of delivery
  - Tissue must be at 4C or on wet ice for <20 hours total (i.e. time from resection to delivery to BWH <20hrs)

## Post Operative Care

- Patients undergoing bone marrow transplant need a 5-7 day period postop for healing before further treatment
- Patients undergoing other therapies may be started on chemotherapy the day after surgery depending on status and the absence of surgical complications. The surgical team will perform a post operative assessment the day after surgery to help with decision making.

### HEPES buffered media:



**M7278** ▶ Sigma-Aldrich

### Minimum Essential Medium Eagle

HEPES Modification, with Earle's salts, 25 mM HEPES and sodium bicarbonate, without L-glutamine, liquid, sterile-filtered, suitable for cell culture

Synonyms: DMEM, MEM

NACRES: NA.75

All Photos (1)

Documents

- SDS
- COO/KOA
- Specification Sheet
- More Documents

SKU	Pack Size	Availability	Price	Quantity
M7278-100ML	100 ML	Available to ship on June 15, 2023 Details...	\$21.00	<input type="text"/> +
M7278-500ML	500 ML	Available to ship on June 15, 2023 Details...	\$51.20	<input type="text"/> +
M7278-6X500ML	6 X 500 ML	Available to ship on June 15, 2023 Details...	\$268.00	<input type="text"/> +

Request a Bulk Order

## **PEDIATRIC SMALL BOWEL FOLLOW THROUGH FOR BOWEL OBSTRUCTION PROTOCOL**

- Communicate with the radiologist that the exam is for small bowel obstruction.
- Allow 24 hours of NGT decompression prior to the start of the SBFT
- Ask radiology if the patient needs another KUB prior to the exam to confirm NGT placement.
- Obtain Omnipaque 300
- Draw up Omnipaque 300 into a Toomey 60ml syringe
- 30 ml for a child less than 5 years old
- 60 ml for a child 5-18 years old
- Bring syringe containing contrast and the portable radiography unit to the patient's bedside.
- Prepare to perform a single view KUB.
- The surgery resident/fellow/APP/attending will inject the contrast via the NGT or g tube.
- Perform KUB immediately after the surgeon injects the contrast medium.
- The NGT or g tube should remain clamped for as long as possible (to be determined by surgical team, but a period of clamping is necessary for the procedure to have effect)
- Ask the nurse to keep the patient upright or RIGHT decubitus for at least 1 hour if possible. This will not be possible in all situations.
- Obtain a single view KUB 12 hours after contrast administration.
- Call radiologist to see whether the exam is complete or if another KUB is needed 24 hours following contrast administration.

\*\*\*Please note that the KUBs may be performed portably or in the radiology suite at the discretion of the surgical team

# UNDESCENDED TESTIS

## TERMINOLOGY

### CRYPTORCHIDISM

Technically a “hidden,” or non-palpable testis; synonym of undescended testis, seen in 5% of full-term male infants at birth. It is bilateral in 1%.

An undescended testis can be located along the normal course of the gubernaculum (normal descent), or be ectopic.

A testis can be undescended (i.e., not in the scrotum), but palpable in the inguinal canal. A testis is impalpable if it cannot be felt on physical exam – either because it is intra-abdominal, or because it is absent.

Retractile testis is a normally descended testis that, at times, retracts up in the inguinal canal, as a result of hyperactive cremasteric muscle. A retractile testis does not require treatment.

## DIAGNOSIS

Normal descent of the testis into the scrotum can still be ongoing at birth, especially in the premature newborn. By 3-6 months, the testis should be in the scrotum. The clinical diagnosis follows this process:

### History

Certain conditions predispose to cryptorchidism: Prader-Willi syndrome, gastroschisis, Opitz syndrome, Noonan syndrome, androgen insensitivity syndrome (AIS) (the latter typically presents as a female phenotype, and is often bilateral).

### Physical exam

#### Empty scrotum

- Was a testis ever felt in the scrotum before? This would suggest a retractile testis
- A hypoplastic (hemi) scrotum suggests true cryptorchidism; a normal (hemi) scrotum may suggest a retractile testis
- In older (school-age) children, secondary cryptorchidism is possible: as the child rapidly grows, a lax gubernaculum can allow a testis to secondarily ascend into the inguinal canal. (This may be more common than previously thought.)
- Palpable testis in the inguinal canal or distal to the external ring (but not in the scrotum): if the testis can be brought in the scrotum and kept there for 10-15 seconds, it may be retractile and observation is sufficient
- Non-palpable testis despite thorough exam: the testis may be high in the inguinal canal, at the internal ring (“peeping testis”), intra-abdominal or absent. Depending on body habitus (esp. in older children), an inguinal testis may be impalpable

### Imaging

- If a testis cannot be palpated, an abdominal (wall) ultrasound may find an inguinal testis, a “peeping testis” (at the internal inguinal ring) or an abdominal testis. If no testis can be found by ultrasound, it raises the suspicion of a vanishing testis or testicular agenesis.
- MRI may be more sensitive than ultrasound in finding an abdominal testis – but surgical exploration would likely still be indicated, regardless of the MRI result

## Medical evaluation

- If genetic or chromosomal anomaly suspected: consider FSH, LH, testosterone levels; Mullerian inhibitory substance (MIS), inhibin B may help diagnose bilateral anorchia if male phenotype with 46,XY karyotype.
- Electrolyte panel, 17-hydroxyprogesterone required in newborn with bilateral cryptorchidism to rule out congenital adrenal hyperplasia

## TREATMENT

### Observation

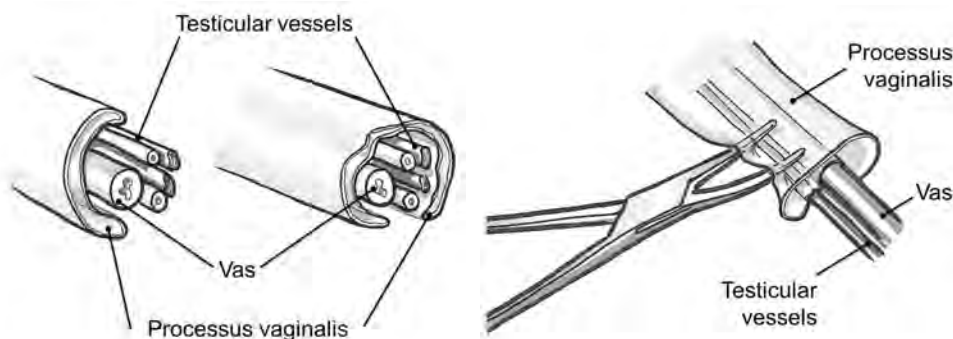
- Appropriate for children under 3-6 months (especially if born prematurely)
- Progression of descent should be monitored in the first 3-9 months
- Arrest of descent by 9-12 months requires intervention

### Primary orchidopexy

- Indicated for testis descent arrested in (distal) inguinal canal by 12 months
- In an older child, even an inguinal testis may be difficult to bring down into the scrotum, and a 2-stage Fowler-Stephens procedure should be considered

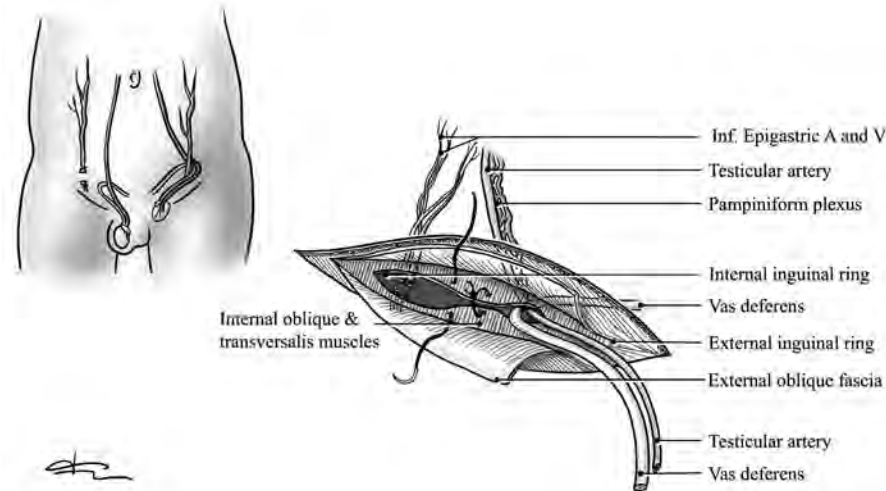
#### Inguinal portion:

- Incision as for an inguinal hernia repair
- Careful dissection of the external inguinal ring, where the testis can be found and freed of distal attachments (gubernaculum)
- External oblique fascia is opened fully
- External spermatic fascia, cremasteric fascia and internal spermatic fascia are divided to expose the testis and epididymis
- Cryptorchidism is usually associated with a patent processus vaginalis (figure 1). The relationship between processus vaginalis, vas deferens and spermatic vessels in cryptorchidism (right) is different than with classic inguinal hernias (left)



- The sac is carefully freed from the vas and vessels (figure 2) and dissected up to the internal ring – then tied off. All cremasteric fibers are divided, to free up the vas and vessels and allow maximal cord length

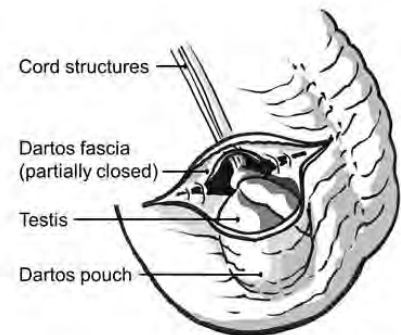
- If the testis reaches the scrotum, it can be brought down. If not, the epigastric vessels may have to be divided to gain length on the cord: This eliminates the internal inguinal ring and requires reconstruction of the floor of the inguinal canal (internal oblique and transversalis muscles) (Prentiss maneuver, figure).



- **If, despite a distal inguinal location, the testis will not reach the scrotum with these maneuvers, it may be best to abandon the procedure early, before devascularization of the vas and testis. A Fowler-Stephens may then be indicated.**

#### Scrotal portion:

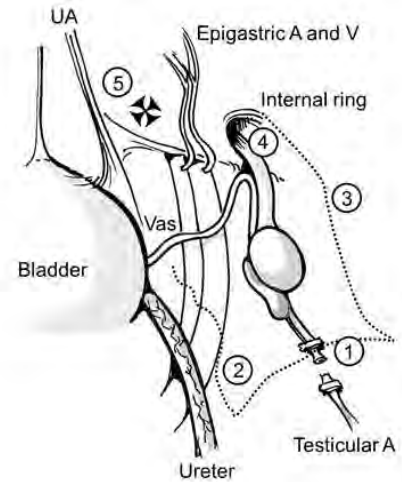
- A transverse skin incision is made (high) in the empty hemiscrotum
- A subcutaneous pocket is bluntly created, between skin (superficially) and Dartos fascia (deep)
- A transverse incision is made in the Dartos, and stay sutures are placed at the two ends
- A long clamp is passed from the scrotal (Dartos) incision into the inguinal incision, and the testis is carefully brought down (check for torsion of the cord!)
- Once the testis lies distal to the Dartos incision, the Dartos fascia is closed around the cord – allowing room for the vas and vessels, but not enough for the testis to come back up (figure 3). The testis may be anchored in its subcutaneous pocket with a vicryl suture
- Both incisions are closed in layers



#### Laparoscopic exploration

- Indicated for non-palpable testis, U/S-proven abdominal testis or “peeping” testis, high inguinal testis in an older child, or absent testis by imaging
- If no testis can be found, the vas deferens is sought, as it crosses the medial umbilical ligament (obliterated umbilical artery) close to the bladder. If no vas can be identified, testicular agenesis is confirmed
- If a vas deferens is present, it is followed distally toward the internal inguinal ring. If it joins the testicular artery and pampiniform plexus and both end blindly, deep to the internal inguinal ring, a vanishing testis can be diagnosed: most often a result of prenatal testicular torsion and necrosis. No further treatment is needed

- If vas and vessels join into the internal inguinal ring, this represents either a vanishing testis or a cryptorchid testis inside the inguinal canal. With preoperative U/S evaluation, an inguinal testis is unlikely to have been missed, but careful inguinal dissection may be indicated (taking care not to damage its blood supply)
- If an abdominal testis or a peeping testis (one that can be brought in or out of the internal inguinal ring with light traction or palpation), a first-stage Fowler-Stephens procedure is indicated
- First stage laparoscopic Fowler-Stephens procedure
- If the decision is made to perform a 2-stage Fowler-Stephens operation, two additional ports are placed (both flanks) – 5 mm diameter
- The testicular artery is identified as high (toward the renal hilum) as possible
- A laparoscopic clip is placed proximally on the testicular artery
- No attempts are made to mobilize the testis, the vas or the vessels
- Second stage laparoscopic Fowler-Stephens procedure
- Typically performed 2-4 months later, to allow secondary vascular ingrowth
- **Abdomen and scrotum are prepped en bloc, and a bladder catheter is placed by the surgeon after draping**
- Same laparoscopic approach as the first stage (same incisions)
- The testis is examined (normal and viable in 80% of cases) and the clipped testicular artery is clipped again, proximally and distally, and divided (figure 4, #1)
- A peritoneal flap is raised as far away as, and parallel to, the artery, in a caudad direction – first medial (#2), then lateral (#3)
- Care is taken to avoid the ureter, medially, and the vas, distally
- Once the flap is raised to the level of the internal ring, the gubernaculum is grasped and stretched out of the inguinal canal (#4). The gubernaculum is divided with cautery **after ascertaining that the vas, which often curls distally to the testis and parallel to the gubernaculum, is safe**
- A scrotal incision is made and a Dartos pouch is dissected, as described above. A long clamp is passed from the scrotal incision to a point lateral to the bladder and the medial umbilical ligament (obliterated umbilical artery, UA) and the epigastric vessels (#5, and X)
- Under constant laparoscopic monitoring, the clamp enters the abdominal cavity, the hole is widened and the testis is brought down into the scrotum (Prentiss maneuver, since the testis now courses medial to the epigastric vessels)
- Alternatively, dilating laparoscopic trocar (7 mm diameter) is introduced over a Veress needle, from the scrotal incision into the abdominal cavity. This has the advantage of minimizing stretch and dilation of the abdominal wall muscles
- The testis is secured in the Dartos pouch as described above



## POSTOPERATIVE CARE

### Immediate postoperative period

- Damage to the testis or the vessels can occur
- Observation for infection, hematoma; relative rest is recommended by some (2 weeks)

### Long-term follow-up

- Testicular atrophy is the most common complication – can be detected at 4-6 months postoperatively
- Success is defined as A) scrotal position of the testis and B) normal volume of the testis
- Success rates are 70-75% for abdominal testes, 85% for canalicular testes and >90% for testes initially located at the external inguinal ring

### Suggested Reading

- Kolon TF, Herndon CDA, Baker LA, Baskin LS, Baxter CG, Cheng EY, Diaz M, Lee PA, Seashore CJ, Tasian GE, Barthold JS. Evaluation and treatment of cryptorchidism: AUA guideline. *J Urol* 2014;192:337-345.
- Petterson A, Richiardi L, Nordenskjold A, Kaijser M, Akre O. Age at surgery for undescended testis and risk of testicular cancer. *N Engl J Med*. 2007;356:1835-41.
- Prentiss RJ, Weickgenant CJ, Moses JJ, Frazier DB. Undescended testis: surgical anatomy of spermatic vessels, spermatic surgical triangles and lateral spermatic ligament. *J Urol*. 1960;83: 686–92.
- Russinko PJ, Siddiq FM, Tackett LD, Caldamone AA. Prescrotal orchidopexy: An Alternative surgical approach for the palpable undescended testis. *J Urol* 2003;170: 2436-8.
- van der Plas E1, Zijp G, Froeling F, de Wilde J, van der Voort L, Hack W. Orchidopexy in late childhood often associated with previously normal testicular position. *Eur J Pediatr Surg*. 2013;23(4):276-82.
- Walsh TJ, Dall'Era MA, Croughan MS et al: Prepubertal orchidopexy for cryptorchidism may be associated with a lower risk of testicular cancer. *J Urol* 2007; 178:1440-6.
- Wenzler DL, Bloom DA, Park JM. What is the rate of spontaneous testicular descent in infants with cryptorchidism? *J Urol*. 2004;171 (2 Pt 1):849–51.
- Wood HM, Elder JS. Cryptorchidism and testicular cancer: Separating fact from fiction. *J Urol* 2009;181:452-61.

## CECOSTOMY TUBES AND INITIATION OF ANTEGRADE ENEMAS

Surgical intervention to help achieve “social continence” may be necessary for patients with poor bowel control, such as those with spina bifida, or with severe and refractory constipation, such as postop anorectal malformation patients. The placement of a cecostomy tube or appendicostomy allows patients to perform large volume antegrade enemas which clean the colon and prevent ongoing daily incontinence.

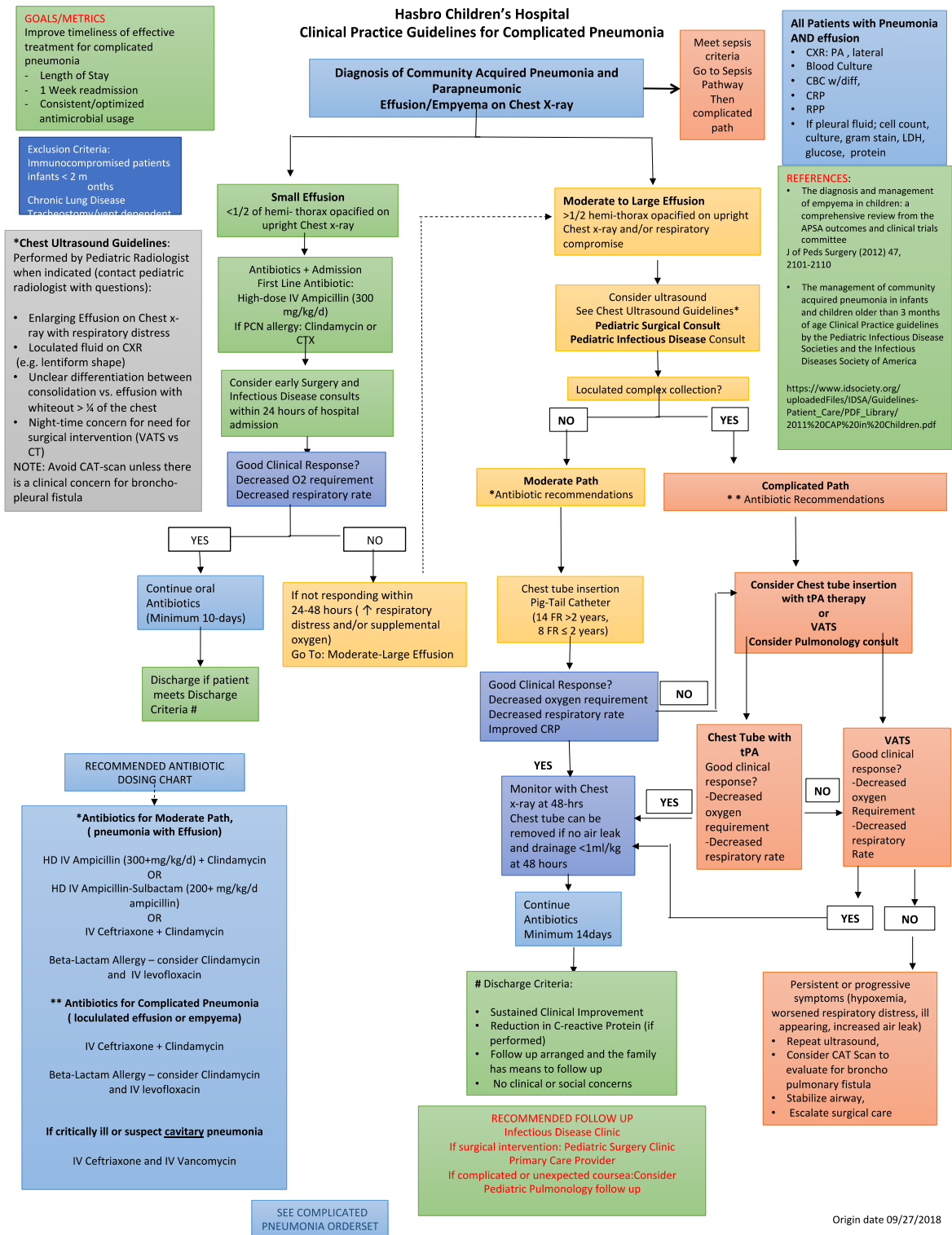
**An appendicostomy** is an access site for antegrade enemas made from the appendix when it is brought to the abdominal wall for intermittent catheterization through an ostomy-like structure; **a cecostomy tube** is placed through the abdominal wall and into the cecum. It stays in place much as a g tube is kept in place for access to the stomach, and like a g tube the track will close if the tube is removed.

Determination of compliance is important preoperatively. For those with refractory constipation, this consists of a 6-month retrograde enema program prior to consideration for surgery. If patients have been successful with the self-irrigations and medications, they can be considered for cecostomy placement. For those with myelomeningocele who have continence issues due to poor neurological and muscular control, retrograde enemas may not be possible and therefore compliance can only be judged indirectly by determining how well they have performed self-catheterizations and how compliant they are with other aspects of their care.

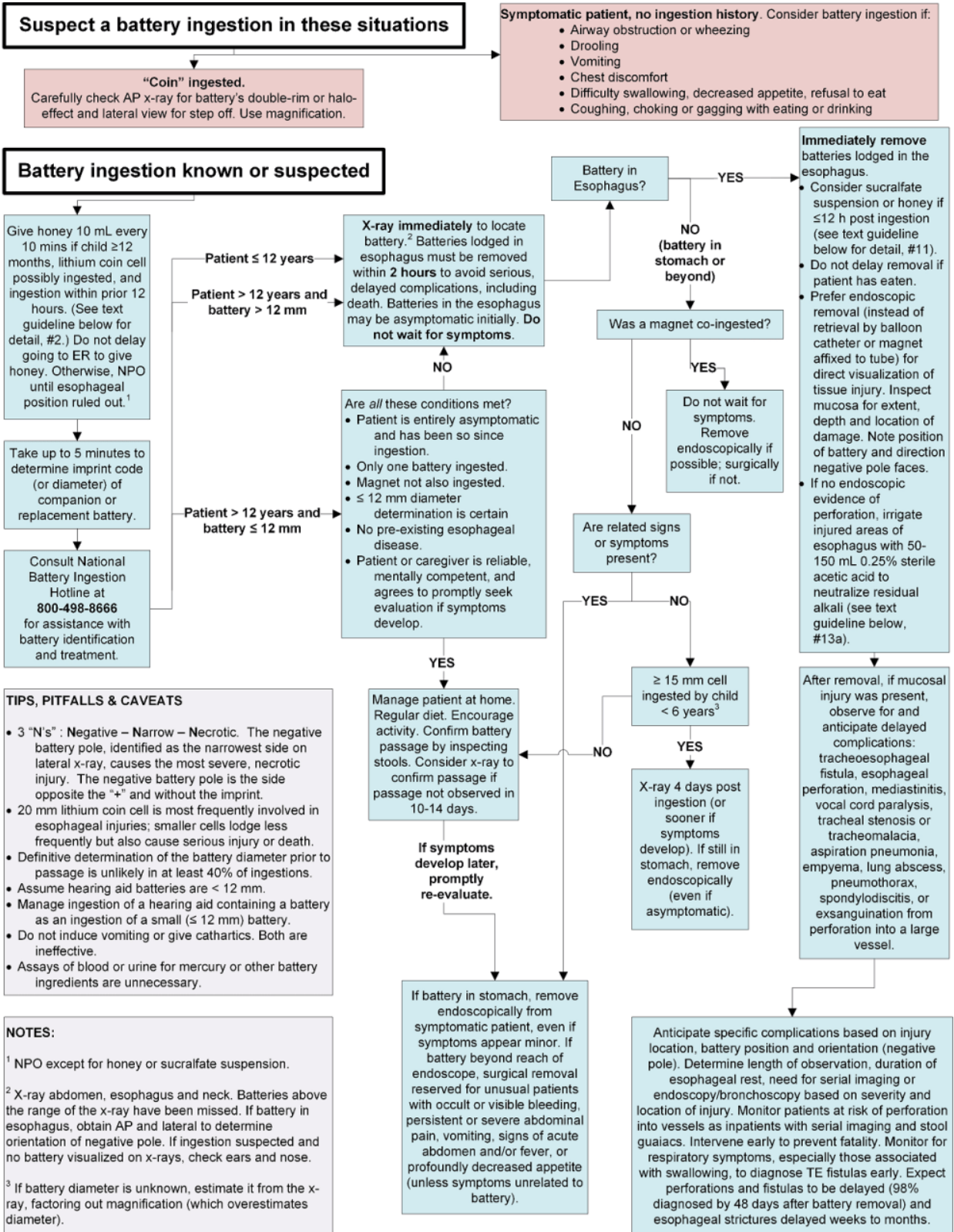
After placement of a cecostomy tube or creation of an appendicostomy in the OR, antegrade enemas and patient/family teaching are begun once bowel function has returned (NGT is out, signs of distal function such as flatus, tolerating diet). Patients are started on 500 cc of normal saline with 30 ml of glycerin over 45 minutes once a day. A small amount of glycerin will initially be provided to the family, but they are responsible for purchasing more once they are discharged. They are encouraged to sit on the toilet for up to one hour. They are then assessed on a case-by-case basis. If no accidents between evacuations occur, then a KUB is obtained to determine fecal load. Depending on the result, the total volume may be increased to 600 cc, again followed by a KUB. If clean, this may be their home dose. If accidents do occur, the amount of glycerin may be increased to 45 cc to help with evacuation.

The goal of initial therapy is no accidents within a 24-hour period and a clean left colon on x-ray. Once achieving these goals, teaching with the family is confirmed and the family is discharged to home. VNA may be helpful, but again should be discussed with the team on a case-by-case basis. An APP will normally call the family a few days after discharge to check on their progress. The patient should be seen in clinic with a KUB one week after discharge.

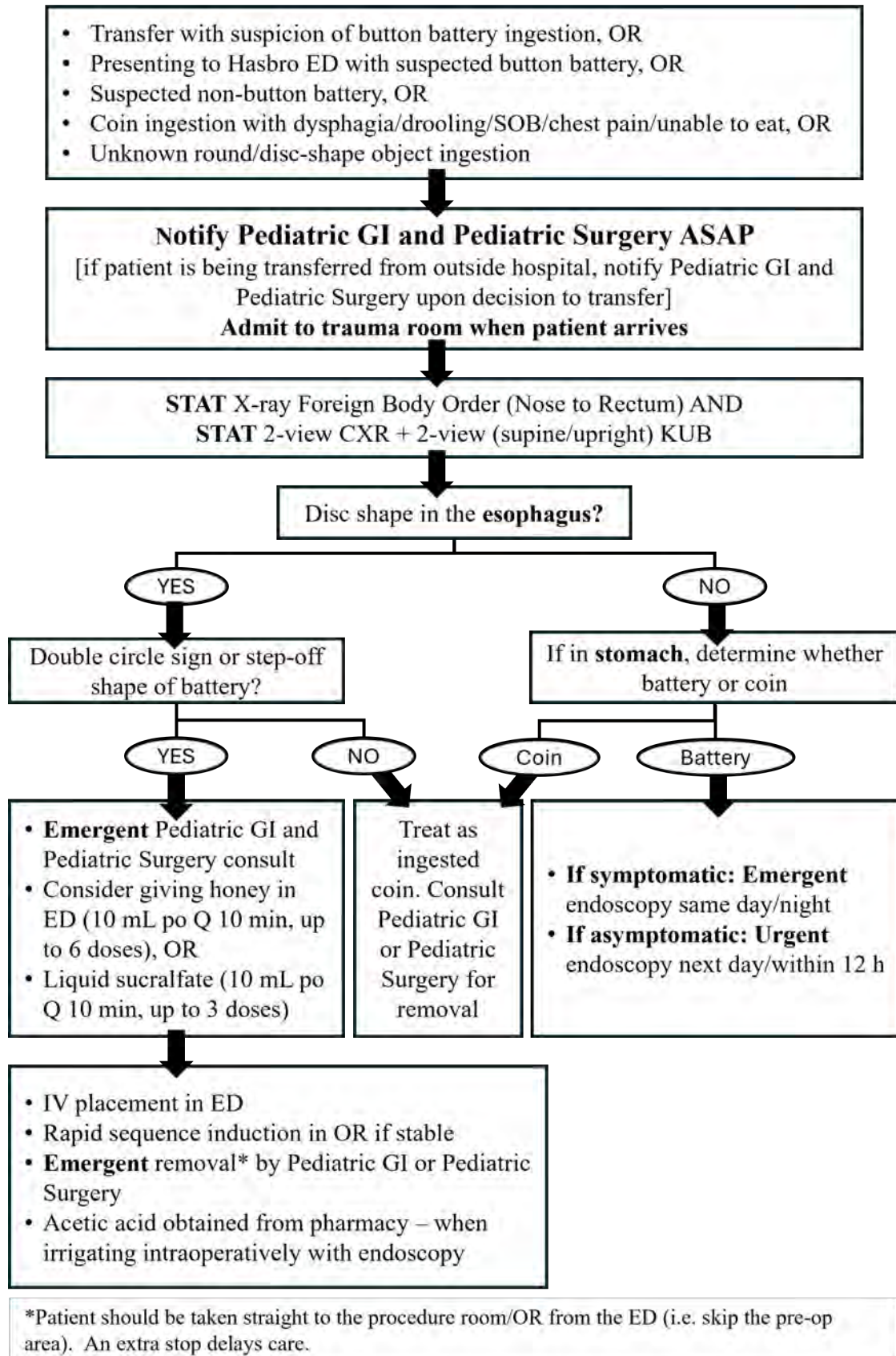
The tube placed initially for cecostomy access is a gastrostomy tube with an inflatable balloon. Like when a g tube is placed in the stomach, the track for this tube needs time to mature. The family should be instructed to call the office and come to the ED if the tube falls out at any time before 6 weeks after surgery, and that only the surgical team replace the tube during this stage of healing.



NOTE: This evidence based guideline was developed for educational purposes and for use at Hasbro Children's Hospital. Decisions about evaluation and treatment are the responsibility of the treating clinician and should always be tailored to the individual clinical circumstances. Contributors: M. Smit, B. Alverson, T. Herliczek, I. Michelow, S. Willis, E. Renaud, L. Polikoff, F. Luks, S. Duffy, L. Mercurio, B. Lee. Direct questions to Sduffy@lifespan.org



## HASBRO BUTTON BATTER INGESTION FLOW CHART



**Addendum for Button/Disc Battery in the Stomach**

- It is rare for a battery that passes through the stomach to require surgical intervention.
  - The battery is unlikely to cause damage in the stomach and could safely be given 24-48 hours to pass out of the stomach IF the patient remains asymptomatic.
  - If the patient remains asymptomatic, but the battery has not passed, a repeat X-ray should be obtained at 48 hours. If the battery remains in the stomach it should be removed.
  - If it has passed out of the stomach and the patient remains asymptomatic, recommend observation at home (looking for battery to pass and/or to see if symptoms develop). If symptoms develop, the patient needs to go to the Emergency Room immediately.
  - Consider repeat radiographs to confirm passage if passage not observed in 10-14 days. Confirming passage may avoid urgent diagnostic intervention for minor symptoms developing later.
- 
- Adapted from <https://www.poison.org/battery/guideline>
  - Anfang RR, Jatana KR, Linn RL, Rhoades K, Fry J, Jacobs IN. pH-neutralizing esophageal irrigations as a novel mitigation strategy for button battery injury. *Laryngoscope*. 2019 Jan;129(1):49-57. doi: 10.1002/lary.27312. Epub 2018 Jun 11. PMID: 29889306.
  - Battery ingestion flow chart 5/2025
  - Developed and reviewed by: Elizabeth Renaud, Prerana Baranwal, Francois Luks, Frank Overly  
May, 2025, \*\*\*

Ed. 06/20/2025